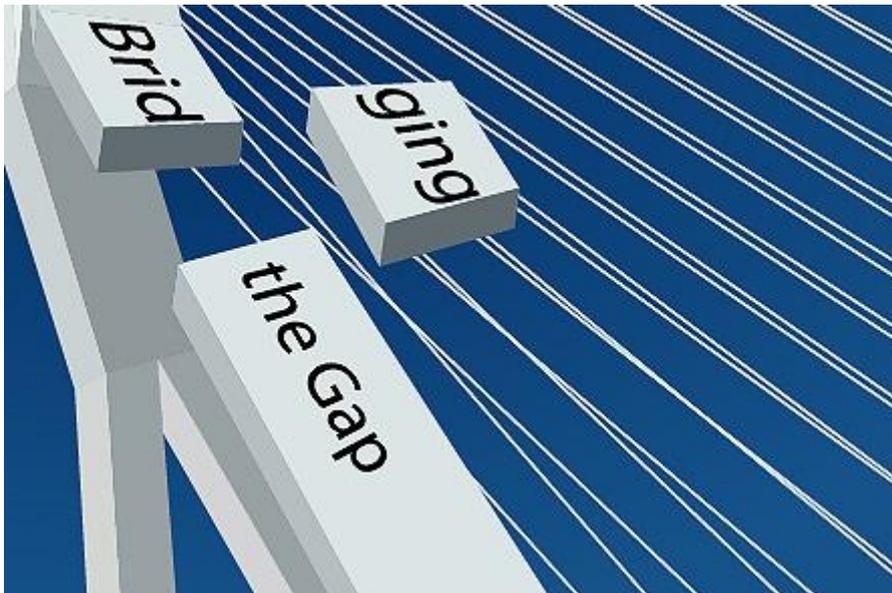


Abstracts

presented during the 3rd International Conference on
Esophageal Atresia



Bridging the Gap

October 2-3, 2014

Rotterdam, The Netherlands

Session 4: Oral presentations (Thursday October 2)

O-01 (16:05 to 16:17)

INFLUENCE OF A VERY LOW BIRTH WEIGHT IN THE SURGICAL MANAGEMENT OF NEWBORN WITH ESOPHAGEAL ATRESIA AND TRACHEO-ESOPHAGEAL FISTULA

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Introduction: Advances in neonatal intensive care have led to a better survival rate in very (<1500g-VLBW) and extremely low birth weight infants (<1000g-ELBW). Some of them are born with esophageal atresia (EA) and tracheoesophageal fistula (TEF).

Aim: To describe the optimal surgical management in this situation.

Methods: Monocentric review of VLBW infants with EA, between 1998 and 2013, treated in our unit. 26 patients (of whom 6 ELBW) were treated. 23 had a TEF. Median gestational age and birth weight were respectively 31 weeks (26-34) and 1245g (600-1470).

Results: The overall survival was 81% with a median follow up of 87 months. For infants with TEF, primary anastomosis was achieved in 7 cases. The 16 remaining had a staged procedure (gastrostomy + TEF ligation and delayed anastomosis). 8 had a gastrostomy prior to ligation, due to pneumoperitoneum or abdominal distension. Patients with isolated EA were treated with delayed anastomosis in 2 cases and coloplasty in one, they all survived. TEF recurrence and esophageal stenosis occurred in 14% and 26%. The survival of infants with a birth weight >1000g was 90%, whereas 50% of ELBW died.

Conclusion: ELBW is a major risk factor in newborn with EA, but babies >1000g have a good prognosis. Early division of the TEF when present is critical; a decompressive gastrostomy may be needed as a first step. Primary esophageal anastomosis may be attempted in stable patients >1000g without pulmonary impairment and/or abdominal distension.

O-02 (16:17 to 16:29)

LATE DEATHS IN PATIENTS WITH OESOPHAGEAL ATRESIA: RESULTS FROM 553 PATIENTS (1980 - 2013)

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Introduction: Whilst most deaths in oesophageal atresia (OA) patients occur in the early postnatal phase, a number of patients will be discharged from hospital only to succumb later in childhood (often unexpectedly).

Aims: To establish the rate and associations of delayed (post-discharge) mortality in our oesophageal atresia cohort.

Methods: Prospectively maintained databases were used to determine the rate and associations of mortality in patients born after 1979. We compared patients with early (in-hospital) mortality (EM) against those with delayed (post-discharge) mortality (DM).

Results: 553 consecutive patients were studied, with 79 (14.3%) deaths identified during the study period. Delayed mortality occurred in 15 (19%). Mean age at death for the DM cohort was 1224 days (100 - 3346 days). The prevalence of the common form of OA (with distal tracheo-oesophageal fistula) was similar between EM (87.5%) and DM (80%) cohorts. Syndromic patients were more common in the EM cohort (39% vs 20%).

The rate of VACTERL was similar between the two cohorts (EM 62.5%, DM 60%). All DM patients had received an oesophageal repair, compared with only 32 (50%) EM patients. The most common causes of DM were sudden unexplained death at home (n = 3), choking episode (n = 3), Fanconi's anaemia (n = 2) and chronic respiratory failure (n = 2).

Conclusion: The rate of DM in patients with OA is higher than previously reported. There are no clear predictors of an increased risk of delayed mortality.

O-03 (16:29 to 16:41)

LONG GAP ESOPHAGEAL ATRESIA : A FRENCH NATIONAL COHORT STUDY FROM 2008 TO 2010

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Aim: To describe current management and mid-term outcomes for patients with Long Gap Esophageal Atresia (LGEA) in France.

Methods: We retrospectively analyzed the whole medical charts of all LGEA cases identified in the French Esophageal Atresia (EA) Registry and managed between 2008 and 2010. We included all infants born with an EA in whom primary anastomosis could not be achieved before 1 month of life because of the length of the gap.

Results: Forty-nine children were included, 37 type I, 5 type II, 6 type III and 1 type IV according to Ladd's classification. The median gestational age and weight at birth were respectively 36 weeks (26.8-41) and 2060 g (550-3740). A surgical gastrostomy was performed in all cases, combined with closure of tracheo-esophageal fistula (n=8), esophagostomy (n=4) or Foker's technique (n=2). Twenty children experienced a complication of whom two died. EA repair was performed either by delayed anastomosis (n=33, 70%) or esophageal replacement (n=14, 30%) at a median age and weight of 87 days (19-454) and 4250 g (2400-8300) respectively. The esophageal replacement techniques were gastric transposition (n=8), gastric tube (n=3), colon interposition (n=2) and Collis-Nissen procedure (n=1). Ninety complications were reported in thirty-eight patients (81%) including 22 anastomotic leaks and 28 anastomotic strictures. At the end of the follow up, the mortality rate was 12%.

Conclusion: Most of the french surgeons management strategies favored a delayed anastomosis for LGEA. Despite a good survival rate of 88%, the morbidity remained high and diverse, highlighting the need for multidisciplinary management.

O-04 (16:41 to 16:53)

THE ROLE OF MAJOR FLAP IN CASE OF LONG GAP ESOPHAGEAL ATRESIA

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Introduction and aim

Long-gap esophageal atresia (LGEA) is a challenging condition to treat even in experienced hands, with high rate of post-operative complications and need for dilatations. Different surgical options, from esophageal traction to substitution, have been proposed to "bridge" the gap.

We review our experience in LGEA, critically exploring the role of major esophageal flap.

Methods

A retrospective analysis of all LGEA treated between January 1995 and December 2013 was performed. Selection of LGEA patients (gap > 3 vertebral bodies) was based on pre and intra-operative esophageal gap measurement. Post-operative outcomes was compared. Statistical analysis was performed with Fisher exact test and Mann-Whitney test, as appropriate ($p < 0.05$ was considered significant)

Results

During the study period we treated 61 patients for LGEA. In 9 cases, major esophageal flap was fashioned to bridge the gap. No differences were found in the outcomes considered. Table summarizes main results.

Conclusions

Long-gap esophageal atresia is frequently encountered in a tertiary referral centre. Extensive preparation of the upper pouch and the creation of major flap represent a feasible approach to bridge the gap, with no increasing rate in post-operative complications and early follow-up.

	Major flap 9	Other LGEA 52	p
Gestation Age	38 (36-39)	38 (35-40)	1
Birth Weight	2700 (2150-3120)	2630 (2200-2980)	0.7
Gap length, cm (IQR)	3.5 (3-4)	3.5 (2.25-4)	0.43
Type A/B vs C/D	6/0 vs 3/0	21/7 vs 24/0	0.7
Cervical esophagostomy (%)	1 (10)	13 (25)	0.7
Age of "definitive" surgery	73 (24.55-183.5)	40 (3.75-141)	0.4
Referral	6 (67)	25 (48)	0.5
Primary anastomosis	7 (78)	36 (69)	0.7
Major leak	1 (11)	6 (16)	1
Re-do esophageal surgery	2 (22)	10 (19)	1
Dilatations, mean (range)	6 (2-7)	4 (2-6)	0.44
Need for esophageal stent (%)	3 (33)	6 (16)	0.1
Segmental resect of stenosis (%)	1 (11)	3 (6)	1
Vocal cord paralyses (%)	0	12 (23)	0.2
Oral aversion (%)	7 (78)	23 (44)	0.08
Antireflux surgery GERD (%)	5 (56)	21 (40)	0.5
Deaths (%)	0	11 (21)	0.2

O-05 (16:53 to 17:05)

LONG-GAP OESOPHAGEAL ATRESIA IS OESOPHAGEAL REPLACEMENT STILL THE BEST OPTION?

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Background/Purpose: Surgical management of long Gap Oesophageal Atresia (LGOA) continues to be controversial . Agreement is that child's own oesophagus is the best. Aim was to review long term results of our management of patients with LGOA+/- fistula.

Methods : Case notes of patients with LGOA +/- fistula managed at our institution between 1989 and 2012 were retrospectively reviewed for type of atresia, sex, weight, GA, associated anomalies, initial management, definitive surgery, and complications.

Results: 22 patients with LGOA +/- fistula (13.4%) were surgically managed. 14 patients (63%) had pure atresia , while 8 had distal fistula with long gap. 12 males and 10 females. Their mean weight was 2568g with a mean GA of 36.6 weeks. Associated anomalies in 45.4% patients. Initial surgery was a feeding gastrostomy in pure atresias and in patients with distal fistula after failed repair. One patient had a failed trial of oesophageal lengthening. Delayed repair was done between days 85 and 147 of life . 5 patients (22.7%) had gastric pullup while 16 patients (72.7%) had delayed primary repair. One patient died a year after delayed repair . One patient had minor leak, while 15 patients (68.1%) developed strictures 4 to 12 weeks post repair that required on average 3.6 sessions of dilation. 6 patients (28.5%) had a fundoplication procedure as part of management of their strictures.

Conclusion: (LGOA) could be treated with delayed primary repair. Strictures and GERD represent the most frequent postoperative problems, additional procedures seem "acceptable"to maintain patient's own esophagus.

O-06 (17:05 to 17:17)

THE MANAGEMENT OF POSTOPERATIVE REFLUX IN CONGENITAL ESOPHAGEAL ATRESIA-TRACHEOESOPHAGEAL FISTULA: A SYSTEMATIC REVIEW

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Introduction: Esophageal atresia (EA), with or without tracheoesophageal fistula (TEF), is associated with postoperative gastroesophageal reflux (GER). GER may exacerbate anastomotic strictures or lead to serious respiratory sequelae.

Purpose: We performed a systematic review of the literature regarding the medical management of GER post EA-TEF repair.

Methods: A comprehensive search was conducted of MEDLINE, EMBASE, CINHAI, CENTRAL, Cochrane Systematic Review Database and grey literature. Full-text screening was performed in duplicate. Included articles reported a primary diagnosis of EA-TEF, a secondary diagnosis of postoperative GER, and primary treatment of GER with anti-reflux medications.

Results: Screening of 2910 articles resulted in 25 included articles (N=1663). Most were single center studies (92%) and the majority were retrospective (76%); there were no randomized control trials (RCT). Fifteen studies named the class of anti-reflux agent used, 3 the duration of therapy and none either the dose prescribed or number of doses. Complications were inconsistently reported. Anti-reflux surgery was performed in 433/1663 (26.0%) patients (23 papers). Average follow-up was 53.2 months (14 studies).

Conclusions: The quality of literature regarding the medical management of GER post EA-TEF repair is poor. There are no well-outlined algorithms for anti-reflux agents, doses, or duration of therapy. Standardized protocols and reliable reporting are necessary for the development of guidelines to decrease postoperative GER in EA-TEF patients

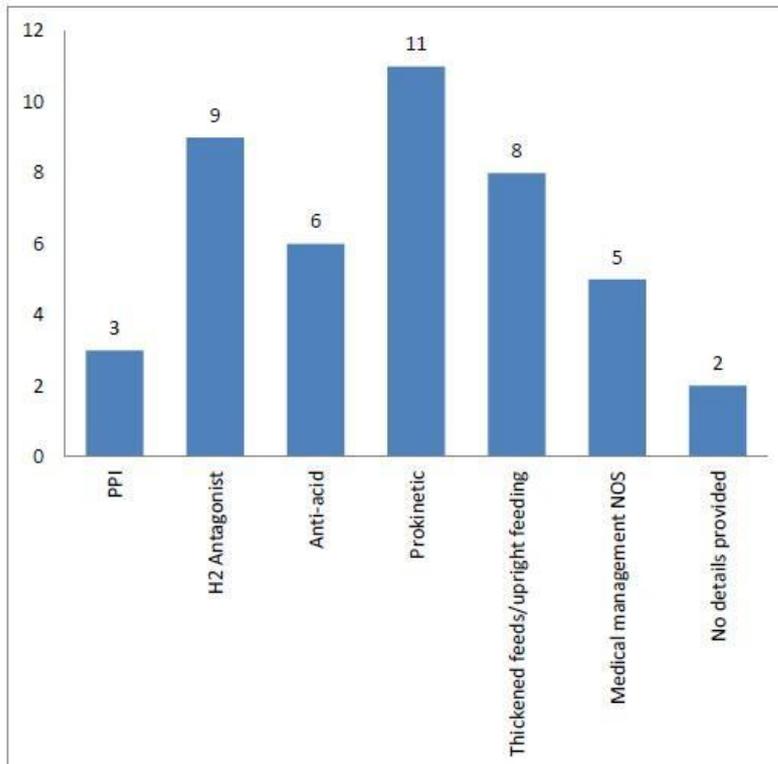


Figure 1: Reporting of medical therapy used in included studies

PPI = proton pump inhibitor

NOS = not otherwise specified, i.e. "medical therapy"

O-07 (17:17 to 17:29)

TRENDS IN UTILISATION OF FUNDOPLICATION IN OESOPHAGEAL ATRESIA: LESSONS FROM 329 PATIENTS (1994 to 2013)

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Introduction: The majority of oesophageal atresia (OA) patients are affected by dysmotility and gastro-oesophageal reflux. In severe cases, particularly those with strictures and/or acute life threatening events, fundoplication may be indicated.

Aims: To establish trends in utilisation of fundoplication in OA over the last 20 years.

Methods Prospectively maintained databases were used to determine the rate and mode of fundoplication for all OA patients within a single centre from 1994-2013. Trends were analysed using Fisher exact or Wilcoxon rank-sum tests to compare 10-year cohorts (1994-2003 vs 2004-2013).

Results: 63 of 329 consecutive OA patients (19%) underwent primary fundoplication, of which 48/63 (76%) were open. Median age at primary fundoplication was 7 (1-44) months. 6/63 (13%) required re-do fundoplication, albeit none since 2001. All re-do operations were open and followed open primary fundoplication. The percentage of OA patients undergoing fundoplication trended down during the study (1994-2003: 36/113, 27% vs 2004-2013: 27/196, 14%; $p=0.004$). Conversely, the proportion of primary fundoplications performed laparoscopically increased (1994-2003: 3/36, 8% vs 2004-2013: 12/27, 44%; $p=0.002$).

Conclusion: Fundoplication remains an integral part of the armamentarium for long-term OA management. These data suggest a more select contemporary utilisation of fundoplication. Laparoscopic fundoplication is gaining favour and will likely become the preferred approach.

Session 5: Dysphagia and dysmotility (Friday October 3)

O-08 (09:45 to 09:57)

FEEDING DISORDERS IN CHILDREN OPERATED ON FOR ISOLATED ESOPHAGEAL ATRESIA

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Introduction: Children operated on for esophageal atresia (EA) often have symptoms concerning acceptance and management of food, cough, choking and vomiting, difficulties in shifting to adult food, long meal time. Despite feeding disorders are frequently reported in these patients (pts), they are scantily investigated.

Aim: To describe feeding disorders observed in a group of children operated on for isolated EA and systematically followed in a dedicated feeding disorders program.

Methods: Pts operated on for isolated EA and followed during 2013 were included in the study. All had video recordings of a mealtime and data on the different feeding/swallowing phases were collected. Phases analysed: general, oral preparation, oral, pharyngeal, esophageal, and gastric. The ability in each phase was graded from 1 (worst) to 4 (best).

Results: During 2013, 21 pts were followed. Male:female ratio: 1:1.1, mean age at the time of the study: 3.4 years (range: 3 months-13 years). All pts had some delay in feeding abilities development (table).

Conclusion: Systematic follow-up of children operated on for EA demonstrates that these patients have disorders in all phases of feeding/swallowing mechanism. Early (since pre-operative period) stimulation of suction and swallowing can contribute to promote and preserve these activities. Parental counselling should focus also on feeding/swallowing disorders, since they may have an impact on the quality of surgical treatment and patients' life.

Clinical manifestation	%
Poor oral intake	47.6
Meal time >45 min	71
Refusal of solid food	70
Prolonged use of feeding bottle	58
Totally parental-assisted feeding	70
Need for distraction	64
Poor management of oral secretions	41
Loss of food during swallowing preparation	62
Bolus stasis in the mouth	87.5
Oral hypersensitivity	100
Chewing disorders (despite adequate age and support)	64
Cough during meal	37
Retching during meal	52
Gastroesophageal reflux	14
Bubbly voice or crying shortly after the meal	33

O-09 (09:57 to 10:09)

TREATMENT OUTCOMES FOR EOSINOPHILIC ESOPHAGITIS IN CHILDREN WITH ESOPHAGEAL ATRESIA

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Introduction: Eosinophilic esophagitis (EoE) has been reported to be more prevalent in esophageal atresia (EA) patients compared to the general pediatric population. To date, little has been reported regarding the treatment outcome in this group of patients.

Aims: To evaluate the responses to EoE treatment in children with EA.

Methods: A retrospective chart review was performed on children with EA and EoE who were diagnosed and treated between January 2000 and September 2013 at the Sydney Children's Hospital. Data collected included details of the patient's treatment, post-treatment endoscopy, symptoms and nutrition.

Results: 23 patients were identified, 4 were excluded due to loss to follow-up. Median age at diagnosis was 26 month (8-103 months) and mean time from diagnosis to last follow-up was 33 months (2-111 months). 8 patients were treated with budesonide slurry, 5 with swallowed fluticasone, 1 with elimination diet and 5 with either budesonide or fluticasone in combination with elimination diet. All patients were on proton-pump inhibitor (PPI) at time of diagnosis which was continued.

Mean peak intraepithelial eosinophil count reduced significantly from 35/HPF (15-80/HPF) to 13/HPF (0-60/HPF) (median time for improvement=22 months) (p=0.001). Of the 4 patients who had furrowing at diagnosis, there was complete resolution in 2 (median time=22 months). Table below shows improvement in histology, symptoms and nutrition with treatment of EoE.

Conclusion: Treatment of EoE in children with EA was found to significantly reduce intraepithelial eosinophil count, symptoms, need for dilatations and improve nutrition.

	Pre-treatment (No. of patients)	Post-treatment (No. of patients)	Median time to Improvement (months)	p-value
Eos/HPF (mean)	35	13	22	<0.001
Furrowing	4	2	22	0.250
Stricture	12	2		0.002
Dilatation (mean number/patient annually)	1.9	1.1		0.004
Dysphagia	13	1		<0.001
Food bolus impaction	1	0		1.000
Dying spells	2	0		0.500
Reflux symptoms	11	2		0.004
Gastrostomy	6	2	12.0	0.125
Weight z-score (mean)	-1.27	-1.14		0.545

Session 6: Barrett esophagus (Friday October 3)

O-10 (11:50 to 12:02)

PREVALENCE OF BARRETT ESOPHAGUS IN ADOLESCENTS AND YOUNG ADULTS WITH ESOPHAGEAL ATRESIA

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Objectives: To study the prevalence of Barrett esophagus (BE) (gastric and/or intestinal metaplasia) in adolescents treated for esophageal atresia (EA).

Methods: This multicentric, prospective study included patients 15 to 19 year-old with medical history of EA. An upper endoscopy with standardized esophageal staged biopsies (at least 12) under general anesthesia was performed. Histological suspicion of metaplasia benefited from a centralized confirmation.

Results: 120 patients aged 16.5 years ($\bar{A}\pm 1.4$) were included. At evaluation, 8% had denutrition, 41% had an anti-reflux surgery performed and 41% had GERD symptoms with only 28% under medical treatment. Esophagitis was found at endoscopy in 34% and confirmed at histology in 67%. Endoscopy suspected BE in 37%, confirmed at histology in 43% (50 gastric and 1 intestinal). No endoscopic nor histological anomalies were found at the anastomosis. No significant relationships were found between BE and clinical symptoms. Multivariate analysis showed that BE was associated to EA without fistula ($p=0.03$; $OR=0,21$ [0,05-0,87]); previous multiple antireflux surgery ($p=0.04$; $OR=3,29$ [1,08-10,07]); BE suspicion at endoscopy ($p<0.001$; $OR=0,04$ [0,01-0,18]); histological esophagitis ($p=0.02$; $OR=0,17$ [0,04-0,73]).

Conclusions: Patients with EA are at high risk of persistent GERD and BE. Long-time systematic follow-up of the esophageal mucosa including multistaged biopsies is therefore required even in asymptomatic patients.

Session 7: (Neuro)developmental aspects (Friday October 3)

O-11 (14:55 to 15:07)

NEONATAL BRAIN OXYGENATION DURING THORACOSCOPIC CORRECTION OF ESOPHAGEAL ATRESIA

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Introduction

Neonates undergoing surgery for congenital anomalies are at risk of adverse neurodevelopmental outcomes, possibly due to perioperative cerebral damage. Near infrared spectroscopy (NIRS) can be applied to measure perioperative regional cerebral oxygen saturation (rScO₂). Little is known about the effects of carbon dioxide (CO₂)-insufflation on rScO₂ during thoracoscopy in neonates.

Aims

To evaluate the effects of CO₂-insufflation on rScO₂ during thoracoscopic esophageal atresia repair.

Methods

Observational study of thoracoscopic esophageal atresia repair, with 5 mmHg CO₂ pressure and flow of 1 L/min. During surgery the mean arterial blood pressure (MAP), arterial saturation, partial pressure of arterial CO₂ (PaCO₂) and the rScO₂ were monitored. Four representative periods of 10 minutes were selected: (T0) during anesthesia before CO₂-insufflation, (T1) after CO₂-insufflation, (T2) at end of CO₂-insufflation and (T3) after desufflation.

Results

14 neonates were eligible for analysis. Duration of anesthesia was 253 minutes [126-500] and of insufflation 156 minutes [74 - 425]. After CO₂-insufflation the MAP showed an increasing trend from mean 43 to 47 mmHg (p=0.08). Arterial saturation decreased from 96.3% to 92.5% (p<0.05). PaCO₂ raised significantly from 47Å±6.9 to 56Å±13 mmHg after CO₂-insufflation and decreased after desufflation to 44Å±7 (p<0.05). rScO₂ remained stable during CO₂-insufflation and after desufflation (rScO₂ (%): 78 [42-85], 72 [36-92], 77 [45-92], 76 [40-93]).

Conclusion

Intrathoracic CO₂-insufflation decreased arterial saturation and increased PaCO₂. Cerebral oxygenation remained stable throughout the whole procedure. Thoracoscopic esophageal atresia repair under insufflation of CO₂ gas with 5 mmHg seems to be safe for preserving cerebral oxygenation in neonates.

O-12 (15:07 to 15:19)

THE EMOTIONAL BURDEN OF ESOPHAGEAL ATRESIA

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Patients with Esophageal Atresia(EA) suffer from chronic gastro-intestinal, respiratory and growth complications, and have an increased risk of esophageal cancer. The emotional burden of this chronic health state on families has been poorly characterized.

Methods:

A bilingual questionnaire was circulated to members of the Association Québécoise de l'Atresie de l'Oesophage during a meeting of parents and patients. It included all four domains of the Parental Experience of Childhood Illness (PECI), as well as demographics and experiential information surrounding the initial NICU visit. PECI data was compared to the original dataset of oncology patients and families using available cumulative statistics, with p<0.05 considered significant.

Results:

Fourteen parents completed the questionnaire, with an average patient age of 6 years. There was an equal gender distribution, and 50% of patients had a type "C" EATEF. Six patients (42%) had an initial hospital stay greater than 3 months. Three parents expressed desire for greater support during their NICU stay. Parents had comparable scores to the original PECI dataset across the domains of Guilt and Worry (p=0.49), Unresolved Sorrow and Anger (p=0.55) and Long Term Uncertainty (p=0.65). The domain of Emotional Resources was superior when compared to the original oncology dataset (p<0.0001).

Conclusion:

While this dataset appears to over-represent complex EA patients, their parents could not be distinguished from parents of pediatric oncology patients across 3 of 4 domains of the PECI, highlighting the significant emotional burden of providing care. Resources to mitigate this burden (multidisciplinary clinics, community meetings) appear warranted.

Poster view (Thursday October 2, 2014 – presenters present: 10:30 to 11:00)

P-01

PROGRESSION TOWARD ORAL FEEDING IN A CHILD WITH LONG GAP EA: A CASE PRESENTATION OF AN INTERDISCIPLINARY APPROACH

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Introduction: Long gap Esophageal Atresia (EA) poses an immediate challenge not only for the surgical and medical teams but to the non-medical team. Aside from possible other malformations or syndromes, long-gap EA signals an immediate and longer term consequence of an inability to feed, which interrupts maternal expectations of nurturing her infant. Repeated surgeries, hospitalizations, dilatations and multiple clinic visits, and the keeping up with multiple medications tend to insidiously wear down maternal energies, and slowly an impatient desire to have the child eat and drink becomes the focus of maternal attention and energies.

Method: Currently there is no universally established protocol for introduction of oral feeding. Each centre develops their own protocol for ensuring eventual oral feeding and drinking skills. In our centre, both the nutritionist and occupational therapist are involved early in the neonatal period to evaluate oral motor skills and to manage exclusive gastrostomy and or jejunostomy. When appetitive issues become apparent, or when mothers become weary of their child's slow progress in obtaining full oral feeding, our feeding psychologist becomes involved.

A case presentation of a child with long-gap EA will highlight the protocol used for stabilising family expectations and establishing full oral feeding by an interdisciplinary team.

Results: Total oral feeding was achieved rapidly by 19 months of age.

Conclusion: The case presentation highlights the importance of a guideline for the introduction of oral feedings in the context of an interdisciplinary team approach to long gap EA.

P-02

DOES PRENATAL DIAGNOSIS MODIFY NEONATAL MANAGEMENT AND EARLY OUTCOME OF CHILDREN WITH ESOPHAGEAL ATRESIA?

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Objective. Our study aimed to (1) evaluate neonatal management and outcome of neonates with either a prenatal or a postnatal diagnosis of EA, and (2) analyze the impact of prenatal diagnosis on outcome according to the type of EA. **Study design.** Population-based study using data from the French National Register for infants with EA born from 2008 to 2010. We compared children with prenatal versus postnatal diagnosis and EA type I and III in regards to prenatal, maternal and neonatal characteristics. We define a composite variable of morbidity (anastomotic esophageal leaks, recurrent fistula, stenosis) and mortality at 1-year.

Results. 469 live births with EA were recorded with a prenatal diagnosis rate of 24.3%. 82.2 % of EA type I were prenatally diagnosed compared to 17.9% of EA type III ($p < 0.001$). Transfer after birth was lower in case of prenatal diagnosis (25.6 vs 82.5%, $p < 0.001$). Delay between birth and first intervention did not significantly differ among groups. Defect size was longer in prenatal diagnosis group (2.61 vs 1.48 cm, $p < 0.001$). The composite variables were higher in prenatal diagnosis subset (44% vs 27.6%, $p = 0.003$) and in EA type I than in type III (58.1% vs 28.3%, $p < 0.001$).

Conclusion. Despite the excellent survival rate of EA, cases with antenatal detection have a higher morbidity related to the EA type (type I and/or long gap). Even it does not modify neonatal management and 1-year outcome, prenatal diagnosis allows antenatal parental counseling and avoids postnatal transfer.

P-03

DELAYED THORACOSCOPIC FOKER LENGTHENING AND KIMURA ADVANCEMENT IN A 1 YEAR OLD

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Aim: To assess the feasibility of a thoracoscopic internal Foker Esophageal Lengthening Technique (FELT) and a Kimura Extrathoracic Esophageal Elongation (EEEEK) to achieve a delayed primary esophageal anastomosis in a 1 year old with long gap esophageal atresia

Methods: An attempted primary oesophageal anastomosis in a new born with esophageal atresia was abandoned after a right thoracotomy and a right neck dissection. A left cervical esophagostomy was formed and feeding by mouth was continued for one year. At one year of age a thoracoscopic internal lengthening of distal esophagus was made to the second intercostal space, using monofilament sutures over radiopaque catheter segments. The cervical esophagostomy was elongated to below the clavicle on the left. Eight weeks later after dissection of the cervical oesophagus a Cluttons dilator was passed from the left neck behind the trachea to enter the apex of the right thorax and out via a thoracoscopic port site. A chest drain was pushed onto the dilator and railroaded into the left neck where sutures on the oesophagus were attached and pulled out of the right chest to maintain traction. A thoracoscopic end to end esophageal anastomosis at the thoracic inlet was performed with 30 braided absorbable suture.

Results: After 48 hours paralysis and 5 days of ventilation, gastrostomy feeds were recommenced. The right intercostal and left neck drains were removed after 8 days. Oral soft diet was commenced 12 days after surgery

Conclusion: Thoracoscopic internal esophageal lengthening and extrathoracic elongation of the esophagus can be delayed

P-04

EARLY COMPLICATIONS AFTER ESOPHAGEAL ATRESIA REPAIR IN A COUNTRY WITHOUT CENTRALIZATION: DATA FROM A GERMAN HEALTH INSURANCE AND COMPARISON WITH THE INTERNATIONAL LITERATURE

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Introduction

The treatment of patients with esophageal atresia (EA) with/without tracheoesophageal fistula (TEF) is not centralized in Germany.

Aim

We hypothesized that the lack of centralization is associated with an unfavorable outcome compared to reports from international centers with a high case-load.

Methods

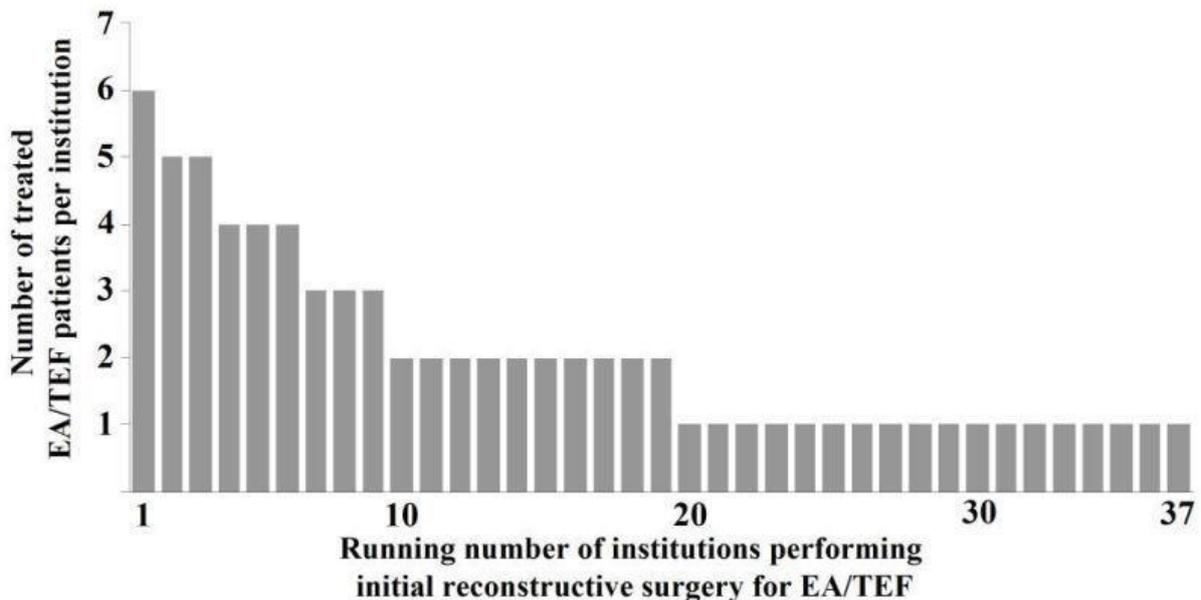
Data from a German health insurance covering ~10% of the population were analyzed. All patients who underwent EA/TEF repair from 01/2007-08/2012 were included. Results were compared to the latest international literature. In addition, various characteristics of the treating surgical institutions (i.e. academic affiliation, number of consultants/beds/preterm infants) were analyzed.

Results

75 EA/TEF patients underwent surgery in 37 pediatric surgical departments [Figure 1]. The incidences of anastomotic leak (3%) and recurrent TEF (7%) were comparable to the literature (both 2-8%). Anastomotic stricture required dilatation in 57% of patients (mean 5.1 ± 5.6 dilatations) comparing unfavourably to most, but not all international reports. 93% of patients were readmitted at least once within one year (mean 3.9 ± 3.1 admissions). The incidence of complications did not correlate with characteristics of the treating institution ($p = 0.05$).

Conclusion

Despite multi-institutional EA/TEF treatment in Germany, operative results were comparable to international reports. A correlation between the complication-rate and characteristics of treating institutions was not identified. Therefore, our unbiased data do not indicate that the lack of centralization is associated with an unfavorable outcome.



Number of patients with EA/TEF who underwent initial reconstructive surgery per institution from 01/2007 to 08/2012.

P-05

A CASE STUDY OF LONG GAP OESOPHAGEAL ATRESIA FOLLOWED TO ADULTHOOD

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Aim: To report a single interesting case study involving the fifteen year follow-up of a child with long gap oesophageal atresia (LGOA)

A 30 week gestation baby was transferred in utero from a peripheral hospital due to concerns over prolonged rupture of membranes. The baby was born in good condition by normal delivery and was noted to have increased secretions and a diagnosis of pure oesophageal atresia was made. A Stamm gastrostomy was made on day 2 of life and a Replogle tube was left in situ. After 6 weeks of aggressive feeding to induce reflux the child proceeded to thoracotomy and a tight anastomosis was performed. He was paralysed for 10 days post-operatively and was ventilated for three weeks. Subsequent contrast showed an anastomotic stricture and gastro-oesophageal reflux (GOR). He underwent serial balloon dilatations and at 7 months of age had an open fundoplication. He made an excellent recovery and was followed up routinely. At the age of four he complained of some dysphagia, but there was no evidence of further stricture and was treated medically for GOR. Further contrast studies over the next decade demonstrated a patent anastomosis and at the age of fourteen he underwent oesophageal manometry and impedance studies. These have interestingly shown no obstruction, but no demonstrable peristaltic waves in the oesophagus. The authors discuss the images and long-term outcome and review the current literature.

P-06

NOVEL TECHNIQUE FOR THE CORRECTION OF SEVERE TRACHEOBRONCHOMALACIA

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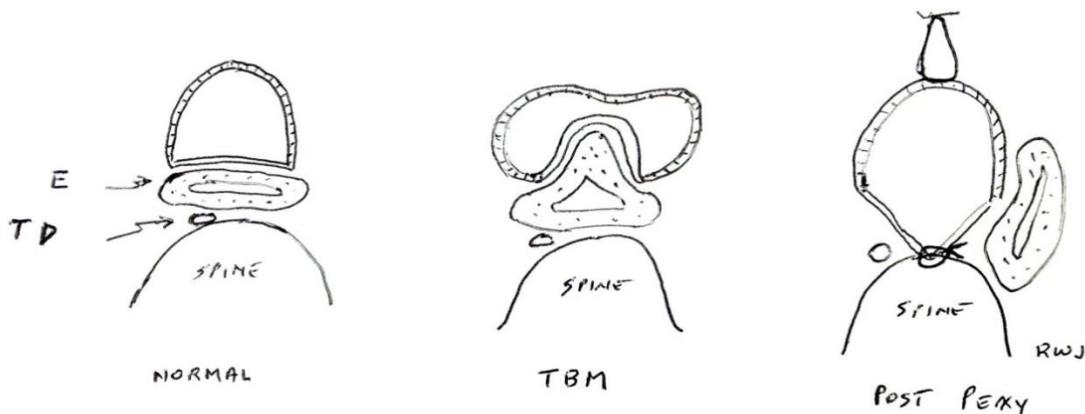
Introduction: Tracheobronchomalacia (TBM) may result in severe, apparent life-threatening events and debilitating respiratory compromise requiring surgery in children.

Aim: Our aim was to describe a series of patients who underwent isolated posterior tracheopexies to treat severe TBM.

Methods: All patients treated at our hospital for TBM from January 2011 to June 2014 were retrospectively reviewed. A previously described classification system was utilized to describe the specific location of malacia. Estimated gestational age, gender, comorbid conditions, endoscopic evaluations, surgical technique, and complications were collected. A novel endoscopic score evaluated severity between preoperative and postoperative exam.

Results: Nineteen underwent posterior tracheopexies (Figure 1). Primary indication was respiratory distress. Thirty-two percent were female. Fifty-three percent were premature. Median gestational age was 37 weeks (range: 31-40 weeks). Seventy-four percent had one-comorbid condition; either cardiac or VACTERL. Eleven-percent had a previously performed operation. Median number of bronchoscopies was 3 bronchoscopies (range 1-10). Median age at operation was 10 months (range: 2-168 months). Median weight was 6.4 kg (range: 3.8-38.8 kg). Operative approach was determined by the surgeon and not randomized; 11% by median sternotomy, 84% by thoracotomy, and 5% utilizing both. No short-term recurrences or complications have occurred. Median decrease in TBM endoscopic severity score was 7 (range: 1-16) in 10 patients.

Conclusion: In this series of patients, an evolution in the manner in which TBM was surgically treated occurred. Given the heterogeneity of this patient population, larger studies are needed to ascertain longer-term outcomes and the validity of the TBM endoscopic score.



Operative approach to isolated posterior tracheopexy

P-07

VASCULAR ANOMALIES ASSOCIATED WITH ESOPHAGEAL ATRESIA AND TRACHEOESOPHAGEAL FISTULA: INCIDENCE, CLINICAL PRESENTATION, DIAGNOSIS AND CONSEQUENCES

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Background: Vascular anomalies may be associated with esophageal atresia (EA) and tracheoesophageal fistula (TEF). Our main objective is to report their incidence in a cohort of EA/TEF patients while describing clinical presentation, diagnosis and consequences. The secondary objective is to determine the diagnostic value of esophagram in the diagnosis of aberrant right subclavian artery (ARSA).

Methods: All patients born with EA/TEF from 2005 to 2013 were studied. Preoperative echocardiography reports, surgical description of primary esophageal repair and esophagram were retrospectively reviewed. Age at diagnosis, discovery mode, clinical presentation and need for surgical correction of the vascular malformation were noted.

Results: 76 of 86 children with EA/TEF were included. Fourteen children (18%) had a vascular malformation. The incidence of right aortic arch (RAA) and ARSA was 6% (5/76) and 12% (9/76) respectively. Respiratory and/or digestive symptoms occurred in 9 of them. Long gap EA and severe cardiac malformations requiring surgery were both significantly associated with vascular anomalies ($p < 0.05$). We reviewed 254 esophagrams; 40% were inconclusive for the detection of vascular anomalies. The diagnosis of vascular malformation was missed in four patients with a long gap EA. The sensitivity of esophagram for the diagnosis of ARSA was 66%, the specificity was 98%, the negative predictive value 95%, and the positive predictive value 85%.

Conclusions: ARSA and RAA have an incidence respectively of 12% and 6% in EA/TEF patients. Echocardiography and esophagram are effective but their sensitivity is not optimal for the diagnosis of ARSA. A CT-angioscan is recommended when esophageal stenting is indicated.

P-08

MULTI-DISCIPLINARY APPROACH TO TRANSITIONING TO ORAL FEEDS FOLLOWING PLACEMENT OF ENTEROSTOMY TUBES IN INFANTS POST REPAIR OF ESOPHAGEAL ATRESIA/TRACHEOESOPHAGEAL FISTULA

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Introduction: Oral feeding post esophageal atresia/tracheoesophageal fistula (EA/TEF) repair can be challenging and require nutrition support via enterostomy tubes (ET) due to strictures, dysphagia and gastroesophageal reflux disease (GERD). This review examines the multi-disciplinary (MultiD) approach used in our institution.

Population: Jan 2011-Apr 2014: 15/36 (42%) EA/TEF repairs required ET, 93% required exclusive ET feeds at discharge. 26% type A; 7% type B; & 67% type C.

Discussion: The MultiD team (occupational therapist, registered dietitian, nurse practitioner & physician) collaborate to support the progression of oral intake through frequent assessments of esophageal patency, oromotor ability, swallowing safety, adequacy of GERD management, & nutritional intervention. Some patients continue to show limited interest in oral intake, hence emphasis is placed on positive oral experiences instead of volume consumed. This helps to establish trust around feeding, which is essential when encouraging progression of oral intake. Family meal times are encouraged to facilitate modeling behaviour and to allow child-led food exploration. When oral liquids are refused, the introduction of developmentally appropriate solid food helps to facilitate oral intake. Throughout, ET feeds are manipulated to support growth while helping to drive hunger/satiety.

Conclusion: A MultiD approach helps to manage the complex factors that hinder exclusive oral feeding in the EA/TEF population by drawing on diverse professional expertise to support a positive feeding environment.

P-09

RECURRENT TRACHEOESOPHAGEAL FISTULA IN ESOPHAGEAL ATRESIA: A MULTICENTER STUDY.

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Recurrent tracheoesophageal fistula (TEF) may occur in 5 to 10% in children born with esophageal atresia (EA) and remains a challenge for both diagnosis and management.

This retrospective multicentre study evaluated, over a 15 years-period, recurrent TEF in children who underwent surgery for EA. Fifty-three patients (27 girls, 84% type III) were identified from 18 centres. Nineteen patients (36%) presented early complications after EA surgery. Twenty-two patients (44%) presented an anastomotic stricture that required endoscopic dilatation.

Recurrent TEF was diagnosed on average 31 months after EA primary repair (range: 8 days - 224 months). Respiratory symptoms revealed recurrent TEF in all cases. Eleven patients (20%) had associated digestive symptoms. An average of 2.7 investigations per child was performed (range: 0-5) including: 43 bronchoscopies (positive in 36/43), 35 esophagograms (positive in 20/35), 27 esophagoscopies (positive in 20/27), 29 combined broncho-esophagoscopies using methylene blue (positive in 26/29).

First line treatment was surgery in 41 cases (85%). Two patients died after surgery. Endoscopic treatment (n=9) was successful in 2 cases but failed in 7 children leading to secondary surgery. A new recurrence of TEF occurred in 15 patients (29%). Finally, closure of recurrent TEF was obtained in 43 cases (84%) after an average of 1.4 intervention per child (range: 1- 4). The mean follow-up was 92 months (range 3-230 months)

TEF can recur a long time after EA repair leading to diagnosis delay. Treatment, mainly surgical, is successful in 85% of the cases.

P-10

HOME HIGH-FLOW NASAL CANNULAE AS A TREATMENT FOR EXTENSIVE TRACHEOMALACIA

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Severe tracheomalacia is a life threatening condition whose management is challenging. Here we describe a case of tracheomalacia treated with long term high-flow cannula. A 39-week gestational age male was noted to have thick secretions, stridor and difficulty breathing at birth. Orogastric tube insertion was unsuccessful and a chest x-ray suggested esophageal atresia. Esophageal atresia type C was confirmed upon surgery. Ligation of the fistula and end to end oesophagus anastomosis were performed. He was extubated within 24h and maintained on Bipap. Esophageal impedancemetry revealed severe gastro-esophageal reflux. The patient underwent a gastrostomy and Nissen fundoplicature. At 2months, he was switched to high-flow nasal cannulae (HFNC) at 7LPM with room air. Attempts to decreased HFNC below 5LPM failed, with increased stridor, SaO₂<90% and PCO₂>65mmHg. Bronchoscopy repeated at 4months showed left main bronchomalacia and severe tracheomalacia with involvement from the cervical trachea to the carina. ChestCT scan showed absence of vascular ring. The patient remained stable on continuous HFNC over the next months. The patient was discharged at 5months on the AIRVO TM 2 humidifier with integrated flow generator at a flow of 5 LPM and room air. At discharge, prolonged pulse oxymetry showed a mean SaO₂ of 97%; PCO₂ was 53mmHg. As the patient continued to evolve well at 9months, HFNC was discontinued during waking hours and finally stopped at the age of 1year. This is the first report of successful treatment of severe tracheomalacia using HFNC. Further studies and follow-up are required before conclusive recommendations can be made.

Poster view (Thursday October 2, 2014 – presenters present: 15:35 to 16:05)

P-12

NUTRITIONAL MORBIDITY FOLLOWING SURGICAL REPAIR IN ESOPHAGEAL ATRESIA/TRACHOESOPHAGEAL FISTULA PATIENTS

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Background: Nutritional morbidity has been reported in infants with esophageal atresia/ trachoesophageal fistula (EA /TEF). Thoracoscopic repair is associated with lower height z-scores in the first year of life and gastro-esophageal reflux disease (GERD) is associated with lower BMI in older children. This study attempts to explore EA-related nutritional morbidity during infancy.

Methods: Single center, retrospective review of repaired EA /TEF infants from Jan. 2011- Mar. 2014 including EA type, co-morbidities, anthropometrics and nutritional interventions. Statistical analysis performed with Stata® 12.0.

Results: Of 35 EA /TEF cases: 11% type A, 3% type B, 77% type C, 9% type E; 46% were premature, 37% had VACTERL associations. Most were repaired by thoracotomy (91%) so type of repair was not examined. Weight z-scores from birth to first follow-up improved ($p=0.03$) with those on promotility drugs (63%) having a trend towards lower weight z-scores at discharge ($p=0.06$) despite receiving similar calories. Overall calorie intake was within estimated energy expenditures (EER). However, calorimetry ($n=13$) revealed resting energy expenditures of $15\pm 26\%$ higher than predicted. Calorie intake did not differ with presence of VACTERL association, whereas calorie intake and prematurity trended towards significance ($p=0.06$). At discharge, 40% required nutrition via enterostomy tube; these patients received significantly more calories than those without tubes ($p<0.05$).

Conclusions: Nutritional morbidities in EA /TEF infants are complex and the current review is limited by the small sample size. Mean EER appears to be within expected norms, but calorimetry suggests a potential for higher than expected caloric requirements. Further assessments are required.

P-13

RATES OF ALLERGIC SENSITIZATION IN CHILDREN WITH ESOPHAGEAL ATRESIA AND EOSINOPHILIC ESOPHAGITIS

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Introduction:

Allergic diseases are increasing in prevalence with 20% of 4 year olds and 40% of 18 year olds sensitized to allergens. Eosinophilic esophagitis (EoE) affects approximately 1:10,000 children of whom 80-90% are sensitized to allergens. We previously reported a higher rate of EoE in patients with esophageal atresia (EA) with 17% of our cohort having EoE.

Aims:

To identify the rate of allergic sensitization in EA patients with EoE.

Methods:

A retrospective review of allergy testing and atopic history in 24 children with EA and EoE.

Results:

Of 24 patients, 12 (50%) patients underwent SPT testing, 8 (33%) underwent both SPT + RAST testing, and 12 (50%) underwent RAST testing. In total, 17 (70%) of 24 patients underwent at least one type of allergy testing, and of these, 15 (88.2%) were found to have a positive allergy test to food and or aero allergens. The most common allergens identified were egg, milk, soy, peanut, and house dust mite. 15 (62.5%) EA patients had an additional atopic history.

Conclusion:

EA patients with EoE have a similar incidence of allergic sensitization to other pediatric EoE patients, and a higher rate than the general population. Proposed mechanisms for allergic sensitization in EA patients include reduced gastric motility resulting in prolonged contact with allergen, contact of allergen with inflamed mucosa resulting in transmucosal sensitization, ubiquitous use of PPIs and limited diet, both of which are associated with increased allergy. We recommend future prospective studies to follow the evolution of atopy in this population.

	All patients	SPT (no. of patients)	RAST (no. of patients)	SPT + RAST (no. of patients)
Total	24	12 (50%)	12 (50%)	8 (32%)
Positive allergy test	15 (62.5%)	9 (37.5%)	10 (41.7%)	8 (32%)
Negative allergy test	9 (37.5%)	3 (12.5%)	2 (8.3%)	0 (0%)

Skin and Serum allergy testing results

P-14

THE MORTALITY OF OESOPHAGEAL ATRESIA: RESULTS FROM 726 PATIENTS (1970 - 2013)

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Introduction: The majority of oesophageal atresia (OA) patients survive through to adulthood. However, a significant minority will die, either early post-natally or later in childhood (often unexpectedly).

Aims: To establish the rate of mortality in our OA cohort, with emphasis upon associated factors that may predict patient demise.

Methods: Prospectively maintained databases were used to determine the rate and associations of mortality in patients born after 1969. Two cohorts (historical [1970 - 89], contemporary [1990 - 2013]) were further analysed.

Results: 726 patients were studied, of which 135 (19%) died during the 44-year period. The mortality rate of the historical cohort (84/330, 25%) was halved in the contemporary cohort (51/396, 13%). This was associated with increased numbers of syndromic patients in the historical cohort (28/84, 33%) (trisomy 21 [n = 6], trisomy 18 [n = 13]) versus the contemporary cohort (11/51, 22%) (trisomy 21 [n = 3], trisomy 18 [n = 4]).

The association between VACTERL and mortality was identical in the cohorts (historical - 31/84 [37%], contemporary - 19/51 [37%]). The decision not to operate was more common in the historical cohort (38/330 [12%] versus 14/396 [4%]) and represented 45% of historical deaths compared with 27% of contemporary deaths.

Conclusion: The mortality rate in patients with OA is higher than previously reported. Historically patients were more often palliated, commonly in the setting of an associated syndrome.

P-15

A SPECIALIZED MULTI-DISCIPLINARY CLINIC FOR CHILDREN WITH ESOPHAGEAL ATRESIA/TRACHEOESOPHAGEAL FISTULA (EA/TEF)

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Introduction

Esophageal atresia with or without tracheoesophageal fistula (EA/TEF) is a congenital foregut anomaly that occurs in less than 1 in 3000 live births (O'Neill's 2004). Innovations and improvements in neonatal resuscitation and management have significantly improved survival of EA/TEF patients. These patients can be very complex due to their neonatal presentation (no antenatal diagnoses, variability in surgical interventions, associated anomalies and morbidities).

Aim

Recognizing the complexity of these patients and the long-term follow up required, a Multi-Disciplinary clinic for EA/TEF patients was initiated at The Hospital for Sick Children in Toronto in 2013. A year following its implementation, we evaluated patient and family satisfaction with a centrally coordinated clinic.

Method

Using patient/family satisfaction surveys developed for out-patient clinics, we evaluated the effectiveness of the one stop clinic approach. Our Multi-Disciplinary clinic involved many services including specialists in pediatric surgery, neonatology, gastroenterology, respiratory and cardiology. We focused on neurodevelopment, feeding and nutritional challenges & respiratory and airway management throughout the lifespan. We also focused on a safe transition from pediatrics to adult care with this complex patient population.

Results/Conclusion

Our evaluation is still in progress however early results indicate that parents are satisfied with the one stop shopping approach to clinic visit and the expertise this clinic is able to provide. We have a family centered approach using collaboration and sharing of expertise to provide essential continuity of care for this patient population.

P-16

THE ESOPHAGEAL ANASTOMOTIC STRICTURE INDEX (EASI) FOR THE MANAGEMENT OF ESOPHAGEAL ATRESIA

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Background

Anastomotic stricture is the most common complication following repair of esophageal atresia. An Esophageal Anastomotic Stricture Index (EASI) based on the post-operative esophagram may identify patients at high risk of stricture formation.

Methods:

Digital images of early post-operative esophagrams of patients undergoing EA repair from 2005-2013 were assessed. Demographics and outcomes including dilations were prospectively collected. Upper (U-EASI) and lower (L-EASI) pouch ratios were generated using stricture diameter divided by maximal respective pouch diameter. Score performances were evaluated with area under the receiver operator curves (AUC) and the Fischer's exact test for single and multiple (>3) dilatations. Inter-rater agreement was evaluated using the intraclass correlation coefficient (ICC).

Results:

Forty-five patients had esophagrams analyzed; 28(62%) required dilatation and 19 received > 3(42%). U-EASI and L-EASI ratios ranged from 0.17-0.70, with L-EASI outperforming the U-EASI as follows: L-EASI AUC: 0.66 for a single dilatation, 0.65 for >3 dilatations; U-EASI AUC: 0.56 for a single dilatation, 0.67 for >3 dilatations. All patients with a L-EASI ratio of 0.30 (n=8) required multiple esophageal dilatations, p=0.006. The inter-rater ICC was 0.87.

Conclusion:

The EASI is a simple, reproducible tool to predict the development and severity of anastomotic stricture after esophageal atresia repair and can direct postoperative surveillance.



ratio 1/2= 0.27 i.e. stricture= 27% of distal esophageal lumen

P-17

INCIDENCE OF MALNUTRITION IN CHILDREN WITH EA-TEF AND THE ROLE OF A MULTIDISCIPLINARY CLINIC IN IMPROVING FEEDING AND NUTRITIONAL OUTCOMES

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Introduction: Children with EA-TEF are known to have feeding and growth difficulties.

Aim: To document the incidence of malnutrition in children with EA-TEF and assess the role of a multidisciplinary EA-TEF clinic in improving feeding and nutrition outcomes.

Methods: Retrospective file audit of patients who attended a multidisciplinary EA-TEF clinic between February 2011 and April 2014. Baseline demographic and nutritional data were collected and compared to data from the last review for those patients who had more than 1 appointment.

Results: 71 patients attended the EA-TEF clinic. 27% were seen once, 49% between 2-3 times and 26% >3 times. There were 30 males. 41% were premature and 27% had an associated anomaly. Median age at initial appointment was 6.5 years (0 -16.8 years). 18 (25%) had a fundoplication and 29 (41%) had gastrostomies of which 10 (14%) were still in use. 68% had dysphagia and 8% were at risk of direct aspiration. At the initial appointment, 73% were seen by a dietician, 58% by a speech pathologist and 97% by a gastroenterologist. Table 1 shows nutrition, growth and constipation at initial and final review. The incidence of constipation reduced from 34% to 13% (P<0.011). Mean weight z score improved from -0.80 (SD 1.25) to -0.74 (SD 1.36), mean length/height z score improved from -0.74 (SD 1.15) to -0.64 (SD 1.26), however this was not significant.

Conclusion: Malnutrition and difficult mealtime behaviours are prevalent in children with EA-TEF. A multidisciplinary EA-TEF clinic can help address some of these issues.

Variables	Initial Review (70)	Final Review (51)
	N (%)	N (%)
Weight z score	N=70	N=51
<1 SD	16 (23)	15 (29)
<2 SD	12 (17)	6 (12)
Height/length z score	N=70	N=50
<1 SD	17 (24)	7 (14)
<2 SD	7 (10)	8 (16)
BMI/weight for length Z score	N=70	N=50
<1 SD	15 (21)	10 (20)
<2 SD	7 (10)	3 (6)
Iron deficient	20/38 (53)	6/19 (32)
Vitamin D deficient	4/28 (14)	2/19 (11)
Constipation	24 (34)	7 (13)
Gastrostomy in use	10 (14)	10 (19)
Feeds increased	2/10 (20)	5/10 (50)
Feeds decreased	5/10 (50)	1/10 (10)
No change	3/10 (30)	3/10 (30)
Closed after initial TOF clinic		2/10 (20)
New gastrostomies from TOF clinic		2 (both have EOE)
Oral supplements	5 (7)	8 (15)
Not accepting age appropriate textures	45/54 (83)	14/46 (30)
Lengthy mealtimes	15/54 (28)	9/46 (20)

Nutrition, growth and constipation in EA-TEF patients at initial and final review

P-18

TWO YEAR NEURODEVELOPMENTAL OUTCOME FOLLOWING NEONATAL REPAIR OF OESOPHAGEAL ATRESIA

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Introduction

The neurodevelopmental trajectory of children with oesophageal atresia (OA) is largely unknown with mixed findings in the limited literature.

Aims

We aimed to determine neurodevelopmental outcome at two years in neonates undergoing OA repair.

Methods

68 neonates underwent repair of OA (2 long-gap) from 2009-2011. Eight (12%) died and 5 interstate patients were not offered follow-up. Nine children were lost to follow-up, 4 declined and 1 is awaiting assessment. 42 two-year-olds were assessed using the Bayley Scales of Infant Development, 3rd edition.

Results

At 2.4 \pm 0.3 years of age 42 children were assessed. Mean birth weight was 2.7 \pm 0.8 kg and gestational age was 37.7 \pm 2.6 weeks. Median age at surgery was 1 (IQR 0-2) day and hospital length of stay (LOS) was 25 (14-38) days. All patients underwent perioperative cranial ultrasound with no significant abnormalities found. Mean cognitive, language and motor composite scores were 102 \pm 13, 101 \pm 16 and 99 \pm 12 respectively. One child was unable to be assessed due to profound deafness and CHARGE Syndrome. Seven (17%) had developmental delay including motor (6), language (3) and cognitive (2) delay. Children with delays were smaller and more premature at birth and had longer LOS than those with normal developmental scores ($p < 0.01$).

Conclusions

Neurodevelopment at two years in survivors of OA is not different from test norms. Intrinsic patient factors are likely important contributors to developmental outcome.

P-19

IMPAIRED PULMONARY FUNCTION AND VENTILATORY LIMITATION IN CHILDREN WITH SUCCESSFULLY REPAIRED OESOPHAGEAL ATRESIA

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Introduction: Significant respiratory morbidity, such as aspiration and recurrent pneumonia occur commonly in young children with oesophageal atresia/tracheo-oesophageal fistula (OA/TOF). Despite clinical improvement with age, lung function may be irreversibly impaired. The role of cardiopulmonary exercise testing (CPET) in the assessment of children with OA/TOF is unknown.

Aim: To assess lung function, exercise capacity and ventilatory reserve in OA/TOF children.

Methods: Prospective evaluation of children \geq 7 years attending a multidisciplinary OA/TOF clinic. Clinical assessment, spirometry and CPET (Bruce Treadmill Protocol) was performed when the child was clinically well.

Results: Eighteen children, aged 7.3-17.9 years, performed spirometry according to ATS guidelines. Fifteen children also performed a maximal CPET. BMI z-score range was -4.3-1.25. Thirty-nine percent experienced chronic cough/wheeze during the last year; 33% had a respiratory admission in the previous year; 11% asthma, 61% had a "TOF" cough, 50% experienced exercise limitation and 33% had undergone an aortopexy. Abnormal spirometry (FVC or FEV₁ < 80%) was noted in 73% and an additional 3 children had distal airway obstruction (FEF_{25-75%} < 80%). Normal exercise capacity was found in 14 children (93%). However ventilation limitation (defined as Ventilator Reserve <20% normal) was observed in 11 (73%). Perceived limitation had a sensitivity of 73%, PPV of 89% and NPV of 57%. Spirometry had a sensitivity of 81%, PPV of 100% and NPV of 67%.

Conclusion: Our preliminary results suggest that perceived exercise intolerance and spirometry are insensitive assessments of ventilatory limitation. A negative history or spirometric evaluation does not exclude diminished breathing reserve.

P-20

EXTREMELY LATE PRESENTATION OF RECURRENT TRACHEOESOPHAGEAL FISTULA

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Recurrent tracheoesophageal fistula is rare. According to the literature, recurrent tracheoesophageal fistula may occur in up to 9% of cases, most often 2-18 months after the initial congenital esophageal atresia repair.

We present here the very rare case of a healthy 42 year old man presenting with a recurrent tracheoesophageal fistula, 42 years after the successful neonatal repair of a congenital esophageal atresia with tracheoesophageal fistula in our own institution, in 1971.

After extensive work-up, the patient was successfully treated by right thoracotomy and fistula resection.

The presenting symptoms, diagnostic work-up and operative treatment will be discussed in detail with an overview of the literature.

This is to our knowledge the longest reported delay for a recurrence of tracheoesophageal fistula after primary successful neonatal repair of congenital esophageal atresia with tracheoesophageal fistula.

P-21

FEASIBILITY AND SAFETY OF SHAM FEEDING IN LONG GAP ESOPHAGEAL ATRESIA

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Introduction: Early introduction of non-nutritive sham feeds in long gap esophageal atresia (LGEA) has been reported in the literature to help prevent long-term oral aversion.

Aim: To report the safety of sham feeds in LGEA; To develop a protocol for routine sham feeds in LGEA.

Methods: A retrospective chart review of LGEA management was performed over a 25-year period. Medical, surgical, and feeding related data were collected. Twenty-nine children were born with LGEA; two were excluded because of early death and reduced past chart. In 2013, following multidisciplinary agreement, a sham feeding protocol was created; two patients benefited from an early version of this protocol.

Results: 27 patients were included in this study. 9 patients were offered sham feeding. Glucose water was most often offered at the time of gavage feeds. Quantities varied from 5 cc to 30 cc. Sham feeds were pursued until time of delayed primary anastomosis in 8 out of 9 patients. There were no medical contra-indications to sham feeding, as there were no complications derived from sham feeding. The two patients who benefited from the protocol seemed to have a decreased tendency to oral aversion while parents greatly appreciated the experience.

Conclusion: Sham feeding appears both feasible and safe in LGEA. This preliminary work has allowed the development of a routine sham feeding protocol at our institution. A long-term prospective study will further help quantify benefits associated with sham feeding.

INTRAOPERATIVE LOCALIZATION OF TYPE C TRACHEOESOPHAGEAL FISTULA USING BRONCHOSCOPIC CANNULATION

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Background: The most common form of tracheoesophageal fistula, type C, is characterized by proximal esophageal atresia and distal fistula location. Surgical repair is standard of care; however, identification of the fistula can prove challenging intraoperatively. At present, there are no studies describing the benefit of fistula localization through intraoperative bronchoscopic cannulation of type C tracheoesophageal fistulas.

Objectives: To demonstrate whether intraoperative localization of Type C tracheoesophageal fistulas through bronchoscopic cannulation is beneficial to the outcome of open surgical repair and to assess for differences in operative time between repair with and without cannulation.

Methods: We performed a retrospective chart review of 12 patients who underwent open surgical repair for type C tracheoesophageal fistula by a single pediatric surgeon, 4 of whom underwent rigid bronchoscopic cannulation by a pediatric otolaryngologist prior to repair. Cannulated and non-cannulated patients were compared for operative times, complications, and recurrences. Independent-samples t-tests were run to determine the difference in operative time.

Results: No statistically significant differences were found in operative time between cannulated and non-cannulated patients. There were no recurrences in either group. No significant complications were encountered. Intraoperative fistula cannulation was found to be of help in documenting the degree of associated tracheomalacia, and to facilitate the pediatric surgeon in localizing the fistula.

Conclusions: Intraoperative bronchoscopic cannulation of Type C tracheoesophageal fistulas directly prior to surgical repair aids in identifying fistula localization and assessing tracheomalacia, thereby facilitating surgical technique without increasing operative time.

Poster view (Friday October 3, 2014 – presenters present 10:10 to 10:45)

P-23

QUALITY OF LIFE IN CHILDREN WITH EOSINOPHILIC ESOPHAGITIS ASSOCIATED WITH ESOPHAGEAL ATRESIA AND TRACHEOESOPHAGEAL FISTULA

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Introduction: Eosinophilic oesophagitis (EoE) has been reported to be more prevalent in esophageal atresia (EA) patients. Both EoE and EA-TEF have been linked to lower Quality of Life (QOL) scores. To date, there have been no studies looking at the QOL in patients with both EoE and EA-TEF.

Aims: To evaluate QOL scores in pediatric patients with EoE and EA-TEF

Methods: Families attending an EA-TEF clinic, whose children had been diagnosed with EoE, were asked to complete the PedsQL Eosinophilic Esophagitis questionnaire. A retrospective audit was performed and patient demographics, prior surgery, stricture, gastroesophageal reflux (GER), histology and EoE treatment data were collected. As there is no validated cut-off value defining 'good' or 'poor' QOL in EoE, a value of 80 was chosen based on a previously published study.

Results: There were 8 children with EoE and EA-TEF, 6 male. Mean age at EoE diagnosis was 2.82 years and mean age at review was 9.54 years. 7/8 patients were on treatment for EoE. The mean QOL score reported by children was 68.35 (range 25-93), whilst the mean score via parent proxy was 63.48/100 (range 17-90). The QOL of the patient who was not on EoE treatment (31.94) was lower than the mean scores on the treated group. Rates of prematurity, GER, strictures, Eosinophils/HPF were higher in children with 'poor' QOL scores. Conversely, rates of fundoplication, gastrostomy and associated anomalies were higher in children with 'good' QOL scores, Table 1

Conclusion: QOL of children with EoE and EA-TEF appears to be low.

	"Good" QOL (score >80) N (%)	"Poor" QOL (score <80) N (%)	P value
All Patients (n=8)	2	6	NS*
Prematurity (n=5)	1 (50%)	4 (66.7%)	NS
GOR (n=6)	1 (50%)	5 (83.3%)	NS
Fundoplication (n=4)	2 (100%)	2 (33.3%)	NS
EoS/HPF (>15) (n=2)	0%	2 (33.3%)	NS
Stricture (requiring >3 dilations) (n=4)	0%	4 (66.7%)	NS
Gastrostomy (n=4)	2 (100%)	2 (33.3%)	NS
Associated anomalies (n=3)	1 (50%)	2 (33.3%)	NS

*NS=not significant

Factors associated with QOL in children with EoE and EA-TEF

NEONATAL MANAGEMENT OF ESOPHAGEAL ATRESIA (EA): A NATIONAL SURVEY OF PRACTICE VARIATIONS IN FRENCH PEDIATRIC SURGERY DEPARTMENTS.

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Introduction Several areas of the management of children with esophageal atresia (EA) remain controversial.

Aim To investigate practice variation in the management of EA in France.

Methods A survey was sent to each of the thirty-eight centers that treat EA in France.

Results Thirty-five centers (92%) responded. If EA has been suspected prenatally, most centers would recommend prenatal MRI (82%). At birth, selective (symptoms or EA suspected prenatally) or systematic diagnostic test with a nasogastric tube was recommended by 30% and 70% of centers respectively. Preoperative echocardiography was almost always recommended (88%). Routine per or preoperative tracheobronchoscopy was recommended by 64% of centers. For EA with distal fistula, 72% of centers recommended thoracotomy, whereas 27% would use thoracoscopy in selected cases. Although 94% of centers recommended placement of a nasogastric tube through the anastomosis, duration and time to initiate enteral or oral feeding were highly diverse. Routine post-operative imaging study of the esophagus was recommended by 64% of centers. The most commonly recommended operation for "long-gap"EA, was delayed anastomosis (88%). If delayed anastomosis was still unachievable, most centers would recommend esophageal replacement (39%) rather than gastric interposition (Spitz) or growth by traction (Foker) (6% and 9% respectively). The preferred technique for esophageal replacement was colonic interposition (73%).

Conclusion This national survey demonstrates that variations exist in the management of EA in France. Variations may represent legitimate decision for varying clinical situation; however it may indicate a lack of clear evidence to guide management or represent heterogeneous standards of care.

RISK FACTORS FOR MORBIDITY AND MORTALITY IN ESOPHAGEAL ATRESIA TYPE III: DATA FROM A POPULATION BASED REGISTER

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Esophageal atresia is a rare disease with a good prognosis especially in lower fistula type. Data about survival and morbidity risk factors are old, issued from retrospective studies and monocentric experiences. The aim of this study was to study risk factors on a large cohort population

Methods

This is a prospective-case populations study including all type III EA recorded in the national register for EA.

Data from pre and postnatal course and associated abnormalities were tested for risk factor of survival and initial morbidity (based on length of initial stay and the absence of nutritional support at first discharge). Logistic and linear regression was used to investigate predictors for survival and early morbidity. All analyses were performed using SAS software version 9.3

Results

Four hundred and nine patients were included. Survival was correlated to associate abnormalities (anorectal $p = 0.01$, CHARGE syndrome $p = 0.001$ and duration of initial length of mechanical ventilation $p = 0.001$). Length of first hospitalization was associated with the existence of prenatal orientation, prematurity, need of nutritional support, difficult anastomosis, and early antireflux procedure surgery ($p = 0.03, <0.001, 0.002, 0.001$ and 0.006 respectively). Full oral feeding at first discharge was negatively correlated to duration of mechanical ventilation, genital and cardiac abnormalities, and positively with standard initial surgery ($p = 0.003, 0.03, 0.01$ and 0.007 respectively).

Discussion:

This first large population based study suggests that malformations and initial intensive care unit stay influence survival, while morbidity is influenced by different independent factors.

P-26

IMPACT OF SUTURE MATERIAL ON THE FATE OF ANASTOMOSIS FOLLOWING ESOPHAGEAL ATRESIA REPAIR

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Introduction: Anastomotic stricture and leak are common complications of esophageal atresia and tracheo-esophageal fistula (EA-TEF) repair. Despite suture material is felt to be important in the healing of the anastomosis, few studies have analysed its impact on the fate of anastomosis.

Aim: To define the impact of suture materials on the outcome of esophageal anastomosis after EA-TEF repair.

Methods: Retrospective study on all patients (pts) treated for EA-TEF between 1995 and 2013. Variables noted included: birth-weight, intra-operative gap (cm), suture material, prevalence of gastro-esophageal reflux, anastomotic stricture (>2 dilatations) and leak, number of esophageal dilatations. Pts were divided based on suture material [absorbable (A) or non-absorbable (NA)] used and compared. Results are prevalence or median (IQ range). X2 or Mann-Whitney test were used; $p < 0.05$ was considered significant.

Results: During the study period, 256 consecutive pts were treated for EA-TEF. Pts with long gap EA-TEF (67), missing data on suture material (10), referred after extensive esophageal surgery (9), H-type TEF (7 pts), or who died shortly after the anastomosis (3) were excluded. The remaining 160 patients form the object of this study (91 group A, 69 group NA). Table shows main findings.

Conclusion: Our data suggest that non-absorbable sutures are associated with lower risk of anastomotic leak in EA-TEF repair. We found no impact of different materials on esophageal stricture.

	Group A (91 pts)	Group NA (69 pts)	p
Birth weight (kg)	2.6 (2.4-3.0)	2.5 (2.2-3.0)	0.5940
Gap (cm)	1 (0-1.5)	1 (1-1.5)	0.1770
GER (%)	19%	21%	1.0000
Stricture (%)	37%	46%	0.2171
Dilatations (#)	1 (0-2)	1 (0-3)	0.1462
Leak (%)	14%	3%	0.0144

P-27

DEVELOPMENT OF INTESTINAL METAPLASIA IN CHILDREN WITH ESOPHAGEAL ATRESIA/TRACHEO-ESOPHAGEAL FISTULA

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Introduction: Patients with esophageal atresia/tracheo-esophageal fistula (EA-TEF) can develop gastric (GM) or intestinal metaplasia (IM) of the esophagus. However IM, which is a major risk factor for adenocarcinoma, has not been well characterized in children.

Aim: Identify and evaluate EA-TEF children with IM.

Methods: Retrospective review of EA-TEF clinic charts of children born and followed at a single center between 1997 and 2012.

Results: Of 137 patients, 2 patients developed IM confirmed on pathology from esophageal biopsies. Both patients had type C EA; patient A was born prematurely at 32 weeks of gestation and suffered perinatal anoxia. Patient B had a VACTERL. While patient A's severely symptomatic reflux (i.e. hematemesis) required both PPI and a Nissen fundoplication, patient B's symptoms were controlled by medication. Both patients developed GM before IM. Patient A developed GM at 6 years of age while patient B first developed GM at 8 years. IM was observed at 11 and 14 years respectively despite proton pump inhibitors or Nissen fundoplication. Interestingly, neither patient had symptomatic reflux at the time of diagnosis of IM.

Conclusion: Patients prospectively and carefully followed in an EA-TEF clinic are prone to develop esophageal IM despite aggressive treatment of GER. This highlights the absolute necessity of a careful endoscopic and histologic surveillance of these patients even in the absence of GER symptoms.

QUALITY OF REPORTING OF THE LITERATURE ON GASTROINTESTINAL REFLUX AFTER REPAIR OF ESOPHAGEAL ATRESIA-TRACHEOESOPHAGEAL FISTULA

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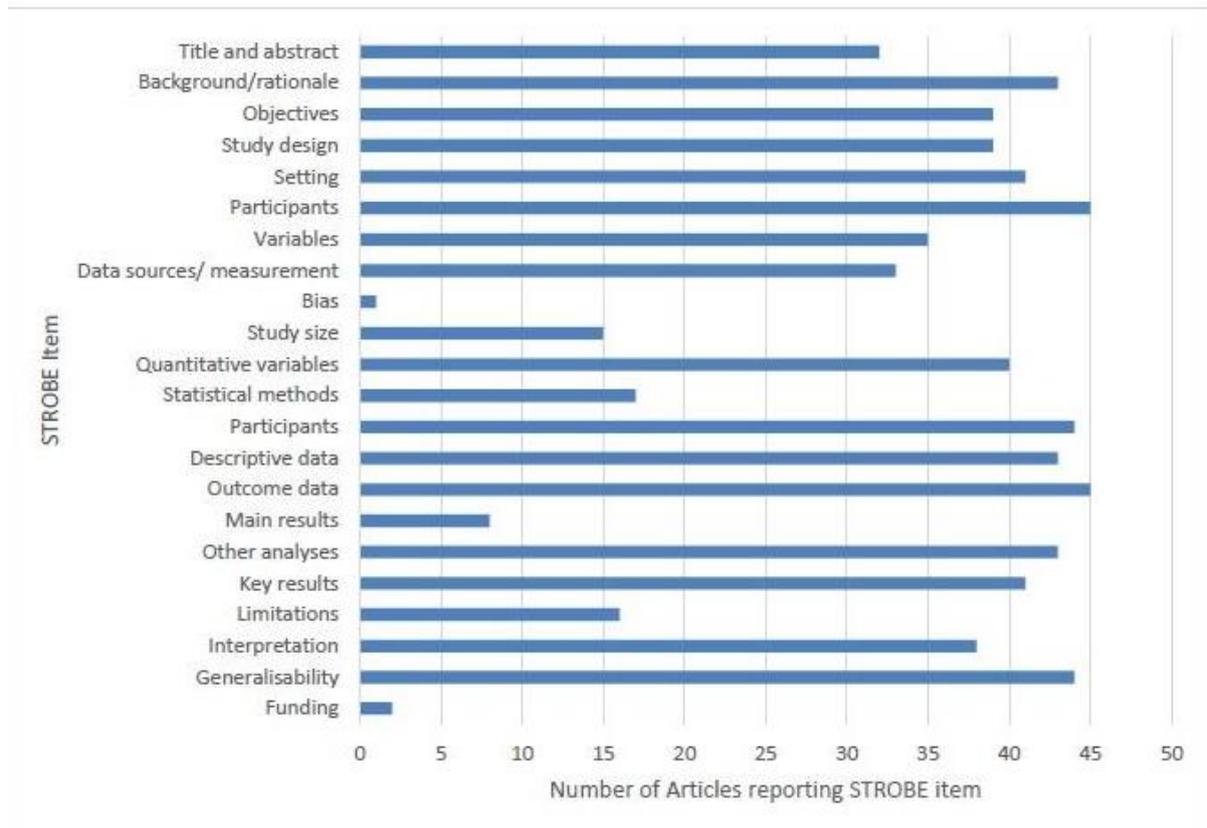
Introduction: There is variation in the management of postoperative gastroesophageal reflux (GER) in esophageal atresia-tracheoesophageal fistula (EA-TEF). Well-reported literature is important for clinical decision-making.

Aim: We assessed the quality of reporting (QOR) of postoperative GER management in EA-TEF.

Methods: A comprehensive search of MEDLINE, EMBASE, CINHAI, CENTRAL databases and grey literature was conducted. Included articles reported a primary diagnosis of EA-TEF, a secondary diagnosis of postoperative GER, and primary treatment of GER with anti-reflux medications. The QOR was assessed using the STrengthening the Reporting of OBservational studies in Epidemiology (STROBE) checklist. An overall quality percentage (OQP) score was calculated.

Results: Retrieval of 2910 articles resulted in 48 relevant articles (N=2592 patients) with an OQP of 48-95% (median=65%). The best reported items were “participants and outcome data”(93.8%), “general results”(91.7%) and “background/descriptive data”(89.6%). Less than 20% of studies provided detailed “main results;”less than 5% of studies reported adequately on “bias”or “funding.”Sample size calculation and study limitations were included in 17 (35.4%) and 16 (33.3%) studies respectively. Follow-up time was inconsistently reported.

Conclusions: Although the overall QOR is moderate using STROBE, important areas are under-reported. Inadequate methodological reporting may lead to inappropriate clinical decisions. Awareness of STROBE emphasizing proper reporting is needed.



Number of articles reporting items on the STROBE checklist

P-29

GROWTH RETARDATION IN PATIENTS WITH ESOPHAGEAL ATRESIA

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Introduction and aim:

Children with esophageal atresia (EA) are at risk for respiratory problems and gastro-esophageal reflux with impaired physical growth. We evaluated growth longitudinally up till 8 years in patients with non-syndromal EA.

Methods

In 128 patients height and weight were measured 438 times at the ages 0.5, 1, 2, 5, and 8 years (N=106, 113, 101, 75 and 43 respectively). Parental height was evaluated as well. Standard deviation scores (SDS) were calculated for height for age and weight for height; and height SDS was corrected for parental height.

Results

Height, parental height corrected SDS (PHC) and weight for height were below the norm (SDS=0) at all time points (all $p < 0.04$). Height, PHC and weight for height did not change significantly with age. All parameters were significantly lower in patients who underwent a Nissen fundoplication ($p = 0.004$).

Conclusion

Our data show that growth is below normal in the first 8 years of life, especially in those treated with a Nissen fundoplication, without significant improvement over time. PHC SDS below normal indicates chronic malnutrition and we recommend evaluation of caloric needs and dietary advice from early age on.

P-30

THE EFFECTS OF CO₂-INSUFFLATION WITH 5 AND 10 MMHG DURING THORACOSCOPY ON CEREBRAL OXYGENATION AND HEMODYNAMICS IN PIGLETS

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Introduction: An increasing percentage of surgical interventions in neonates is performed by minimal invasive techniques. Recently, concerns have been raised regarding a decrease of cerebral oxygenation in neonates during thoracoscopy.

Aim: To evaluate the effect of CO₂-insufflation with 5 and 10 mmHg on cerebral oxygenation and hemodynamics in neonates.

Methods: Piglets were anaesthetized, intubated, ventilated and surgically prepared. CO₂-insufflation was done with 5 or 10 mmHg CO₂ during one hour. Arterial saturation (SaO₂), heart rate (HR), mean arterial blood pressure (MABP) and cerebral oxygenation (rScO₂) were monitored. cFTOE, an estimator of cerebral oxygen extraction ($(SaO_2 - rScO_2) / SaO_2$), was calculated. Arterial blood gases were drawn every 15': pre (T0), during (T1-T4) and after CO₂-insufflation (T5).

Results: Two piglets in the 10mmHg group needed acute resuscitation and none in the 5mmHg group. Piglets insufflated with 5 mmHg CO₂ show a stable saturation, HR and MABP during procedure. The pCO₂ (mmHg) increased from 36 Å±4 at T0 to 70 Å±19 at T4. The rScO₂ (%) increased from T0 42 Å±3 to 57 Å±1 at T5. Thoracoscopy with 10 mmHg gave a decrease in MABP from 84 Å±8 at T0 to 54 Å±21 at T3. HR increased from T0 152 Å±18 to 218 Å±9 at T3. The rScO₂ (%) showed an increase from 37 Å±4 at T0 to T5 50 Å±5. The cFTOE was higher in 10mmHg, which suggests a lower cerebral perfusion in comparison to 5 mmHg. **Conclusion:** Insufflation of CO₂ during thoracoscopy with 10 mmHg caused more severe hemodynamic instability and seems to be related with a decrease of cerebral perfusion as represented by a higher oxygen extraction. CO₂-insufflation of 5 mmHg for thoracoscopy seems to have no adverse effects on cerebral oxygenation.

P-31

ESOPHAGEAL REPLACEMENT BY A TISSUE ENGINEERED SUBSTITUTE IN A PORCINE MODEL

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Introduction: Oesophageal replacement by the colon or the stomach for malignant and benign oesophageal diseases exposes to significant morbidity and mortality.

Aim: In that setting, tissue engineering appears a seductive alternative.

Material and methods: In a porcine model, the success of the 5 cm-long circumferential replacement of the cervical oesophagus by a substitute made of an acellular matrix (SIS) seeded with autologous skeletal myoblast coupled with a human amniotic membrane seeded with autologous oral epithelial cells was assessed. The substitute was matured 3 weeks in the great omentum before esophageal replacement. Eighteen minipigs, (group A (substitute with esophageal endoprosthesis, n=6), group B (substitute alone, n=6), group C (endoprosthesis alone, n=6) were used. The esophageal endoprosthesis was removed endoscopically at 6 months. Animals were sacrificed sequentially over a 12 month-period. Clinical, endoscopic, radiological and histological outcomes were analysed.

Results: All animals of group B and C died during the first two months because of refractory esophageal stenosis or endoprosthesis extrusion. Nutritional autonomy without endoprosthesis was observed in all animals of group A surviving more than 6 months (n=3). A phenotype similar to native esophagus, consisting in a mature epithelium, submucosal glands and a circular muscular layer, was observed after 9 months.

Conclusion: In this model, the circumferential replacement of the cervical esophagus by a hybrid substitute composed of two different matrix and cellular cell types allowed, under the temporary cover of an esophageal endoprosthesis, the recovering of nutritional autonomy and tissue remodeling toward an esophageal phenotype

P-32

THE DEVELOPMENT OF A MULTIDISCIPLINARY TEAM TO EVALUATE EARLY DEVELOPMENTAL OUTCOMES OF INFANTS WITH TOF/OA

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Aims

To determine enrolment in the clinic and subsequent developmental outcome of a cohort of infants with TOF/OA over the last three years.

Methods

Infants admitted with the diagnosis of TOF/OA were identified from the neonatal unit database and developmental outcomes from the clinic database. Infants were assessed using the Bayley-III and reasons for non-assessment were recorded.

Results

Of 29 possible infants, 18 were enrolled in the clinics, with 16 infants completing developmental assessments. Developmental assessments showed that most infants were performing within the average range on each of the subtests, with the greatest delay found in gross motor with 33% scoring below average. Of the 13 infants who were not assessed, the majority were for reasons such as parental refusal, non-attendance, infants enrolled in other neurodevelopmental clinics and infants from overseas.

Conclusions

NSW is a very large state, of over 800,000 square kilometres and has a population of over 6.7 million. The distance which parents have to travel makes standardised follow-up difficult. We have recently commenced multi-disciplinary clinics for TOF/OA infants where in the one clinic they see a surgeon and a respiratory paediatrician. It is planned to combine this with neurodevelopmental assessment to provide a one visit clinic assessment which will benefit the child and parents. Although five infants were not enrolled, additional strategies to prevent this have resulted in no infants being missed since the beginning of 2013.

	Below average n (%)	Average n (%)	Above average n (%)
Cognition n=16	1 (6)	12 (75)	3 (19)
*Receptive language n=15	2 (13)	12 (80)	1 (6.7)
*Expressive language n=15	2 (13)	13 (87)	0
Fine Motor n=16	1 (6)	13 (81)	2 (12.5)
*Gross motor n=15	5 (33)	10 (67)	0

*1 not complete

Developmental outcomes

BARRETT'S ESOPHAGUS AND EOSINOPHILIC ESOPHAGITIS IN A YOUNG PATIENT WITH ESOPHAGEAL ATRESIA

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Barrett's (BE) is considered a chronic, long term complication of esophageal atresia (EA). Eosinophilic esophagitis (EoE) has only rarely been reported in EA patients. We describe a young patient who was discovered to have both BE and EoE at the early age of 4 years, post EA repair.

Our patient is a 4 year old girl who was born with long gap, Type A EA and underwent a delayed primary repair (Foker technique) when she was 3 months old. No fundoplication was performed. She was non compliant to anti reflux medication. At age 4 she was referred for worsening dysphagia and food impaction despite a normal contrast study. First endoscopy showed esophageal furrowing and exudates. EoE was confirmed on biopsies from the proximal and distal esophagus. She was commenced on Omeprazole for acid suppression therapy and swallowed Budesonide slurry. A 3 month follow up endoscopy revealed improvement in her EoE but Barrett's mucosa was noted in the distal esophagus. This had not been seen at initial endoscopy. Histology confirmed intestinal metaplasia with no dysplasia.

To date, our patient is the youngest EA patient in reported literature to have developed Barrett's changes and the first reported EA patient with both EoE and BE. The discovery of BE in our young patient suggests that early and regular endoscopic surveillance is vital from a young age. The concomitant presence of EoE further raises the possibility that EoE may be an independent risk factor for the development of BE.

Poster view (Friday October 3, 2014 – presenters present 15:20 to 15:50)

P-34

MOTOR PERFORMANCE AND EXERCISE CAPACITY IN CHILDREN BORN WITH ESOPHAGEAL ATRESIA: A LONGITUDINAL EVALUATION

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Introduction With increasing survival rates of children with esophageal atresia (EA) attention is shifted from mortality towards morbidity. We previously reported motor function problems at 5 years in 34% of EA survivors (n=29).

Aim To evaluate longitudinally motor performance and exercise capacity in 5- and 8-year old EA patients.

Methods Children born with EA participated in a prospective longitudinal follow-up program. Motor performance was evaluated using the Movement Assessment Battery for Children (MABC) and exercise capacity by the Bruce treadmill protocol. We used z-scores for longitudinal evaluation.

Results We assessed 53 and 43 children at 5 and 8 years, respectively; 7.5% patients had type A and 92.5% had type C EA.

Motor performance: at 5 years, 68.0% scored within normal range, 20.0% was classified as borderline and 12.0% had a definite motor problem; mainly ball- and balance skills. At 8 years: 68.3% normal; 12.2% borderline and 19.5% definite motor problem (mainly ball-skills). At five, the mean z-score MABC was significantly below normal: -0.70; $p < .001$, without significant change over time (mean difference: +0.21, $p = .20$).

Exercise capacity: mean (SD) z-scores endurance time were -0.6 (1.1) at 5 and -1.0 (1.0) at 8 years, both significantly below zero ($p < .01$).

Conclusions Gross motor performance and exercise capacity were significantly impaired at 5 years in EA patients and had not improved at 8 years. Intervention should aim at both domains.

P-35

GASTRIC METAPLASIA IN PATIENTS WITH ESOPHAGEAL ATRESIA/TRACHEO-ESOPHAGEAL FISTULA (EA-TEF) AGED LESS THAN 10 YEARS.

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Introduction: EA-TEF patients can develop Barrett's esophagus (BE) which requires careful follow-up. Chronic reflux is associated with BE in adults, but incidence and risk factors for developing BE in EA-TEF are unknown.

Aim: To determine 1- The incidence of gastric metaplasia (GM) and 2- Factors associated with GM in EA-TEF patients.

Methods: Retrospective review of EA-TEF clinic charts of 77 children born 2004-2012. Death (n=6), children followed elsewhere (n=6) or patients who had had no esophageal biopsy (n=13) were excluded.

Results: Of 52 patients evaluated (mean follow-up 6.5 y; range 1.5-10), 6 (12%) had an endoscopy suspected BE with GM confirmed at histology. No patients developed intestinal metaplasia. There was no difference in atresia type nor associated malformations between patients with GM with those without GM (NM). The anastomotic leak rate was similar (28% vs 33%, NM vs GM). 41% of NM patients developed anastomotic strictures requiring a mean (range) of 3.3 (1-8) dilatations, vs 66% of GM patients (7 (3-17) dilatations). No patients with GM tolerated discontinuation of PPI therapy versus 53% of NM patients ($p < 0.01$, χ^2 test). Higher rates of esophagitis were observed on endoscopy or histology in patients with GM (83% vs 37%; $p < 0.01$).

Conclusion: 12% of EA-TEF patients < 10 years followed in a single center EA-TEF clinic developed esophageal GM. These patients had more severe gastroesophageal reflux. This underscores the need of a close endoscopic follow-up of these children.

QUALITY OF LIFE, FAMILY IMPACT AND PARENTAL SATISFACTION WITH MULTIDISCIPLINARY CARE IN CHILDREN WITH OESOPHAGEAL ATRESIA

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Introduction: Significant morbidity, including gastro-oesophageal reflux disease (GORD) and recurrent lower respiratory tract infections (LRTIs) occur in children with oesophageal atresia (OA). Quality of life (QOL) may be impaired and negatively impact on family functioning. Multidisciplinary care may improve health outcomes and increase parental satisfaction.

Aim: To assess QOL, parental impact and healthcare satisfaction scores in paediatric OA.

Methods: Prospective evaluation of children/families attending a multidisciplinary OA clinic. Parental impact, QOL and healthcare satisfaction were assessed using the PedsQL Family Impact Module 2.0, PedsQL 4.0 Paediatric Quality of Life Inventory and PedsQL Healthcare Satisfaction Generic Module 3.0 (PedsQLHCS). Responses were grouped according to presence of associated congenital anomalies.

Results: Fifteen children, 2.8-17.9 years, mean 7.8 (4.5), gestation (GA) 28-40 weeks, mean 35.8 (3.5), 60% with associated cardiac, skeletal and other abnormalities, and their families completed the questionnaires. Twelve (80%) had type C OA. All children had significant past GORD; 60% were undergoing current treatment. Eighty percent experienced LRTIs in the past 2 years. Parents of children with additional anomalies scored lower in the "Technical Skills" domain of the PedsQLHCS questionnaire, median 75 [71.2 -100] out 100, compared to perfect scores in parents of children with isolated OA, $p=0.036$. Furthermore, they scored lower in "Emotional Needs", median 70.8 [52.1 - 95.3] vs. 100 [93.8-100], $p=0.043$.

Conclusion: Our preliminary results suggest that parents of OA children with additional congenital anomalies perceive lower staff responsiveness to the psychosocial needs of their children. Children with complicated OA require holistic multidisciplinary care.

*Mann-Whitney U

P-37

THE EVALUATION OF DEGLUTITION AND DYSPHAGIA AFTER REPAIR OF ESOPHAGEAL ATRESIA AND TRACHEOESOPHAGEAL FISTULA

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Aim: To investigate the clinical problems and dysphagia in follow-up of the cases operated for esophageal atresia and tracheoesophageal fistula (EA-TEF), and to evaluate the videofluoroscopy (VF) findings of deglutition.

Methods: Patients with EA-TEF repair were evaluated for type of anomaly, operative procedure, postoperative complications, deglutitive and respiratory symptoms, gastroesophageal reflux disease (GERD), dysphagia score and VF findings of oral, pharyngeal and esophageal phases of deglutition.

Results: Twenty nine cases with a median age of 58.8 months (4-120 months) were included. Male female ratio was 14:15. The most common type of anomaly was the Gross type C (n:24, 82,8%), and the others were type A (n:2), D (n:2) and E (n:1). EA repairs were performed with primary (n:24, 82,2%) and delayed (n:5, 17,2%) anastomosis. Postoperative complications including anastomosis stricture (n:10), leak (n:2) and recurrent fistula (n:1) were managed by dilatation, conservative approach and repair of the fistula, respectively. Eleven of the cases had recurrent pneumonia. The diagnosis of GERD was made by contrast graphy (n:15) and pH monitorization (n:9). Dysphagia for liquids and solids were absent (n:24, n:19), improved (n:3, n:5), or going on (n:2, n:2) respectively. Mean dysphagia score was 3,48 (min:0-max:27), and videofluoroscopic findings were listed in Table 1.

Conclusions: The cases with repair of EA-TEF may reveal deglutitive and respiratory symptoms in follow-up, necessitating certain investigations. The deglutition is functionally evaluated with VF, and while the disorders of oral and pharyngeal phases are less frequent and prominent, the esophageal phase reveals disorders with higher incidence and severity.

VF findings	Normal (n)	Mild (n)	Moderate (n)	Severe (n)
Oral phase				
Lip closure	27	1	1	0
Tongue elevation	26	1	2	0
Tongue retraction	26	2	1	0
Oral retention	26	2	1	0
Pharyngeal phase				
Delay in deglutitive reflex	23	4	2	0
Touch of root of tongue to pharynx	26	0	3	0
Velopharyngeal closure	26	0	1	0
Hyopharyngeal elevation	24	2	1	2
Closure of airway	24	1	1	3
Vallecular retention	26	2	1	0
Pharyngeal retention	28	1	0	0
Retention in pyriform sinus	25	3	1	0
Aspiration	26	3	0	0
Silent aspiration	26	3	0	0
Esophageal phase				
Opening of UES	23	5	1	0
UES retention	11	10	5	3
Esophageal back-flow	4	4	7	14
Motility problem	2	1	7	10
Esophageal retention	3	3	11	12
LES dysfunction	6	13	8	2

VF findings of patients with EA-TEF

P-38

RISK FACTORS FOR EOSINOPHILIC ESOPHAGITIS IN CHILDREN WITH ESOPHAGEAL ATRESIA.

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Aim: Eosinophilic Esophagitis (EoE) is reported in esophageal atresia (EA) but risk factors are unknown.

Methods: 31 children (16M/15F) with EA underwent diagnostic upper endoscopy (1y to 16y, mean age 6y) and records were reviewed for risk factors including estimated gestational age (EGA), mode of delivery, anastomotic dilation, family history of atopy, neonatal milk type, inhaled steroid use, tracheomalacia and aspiration pneumonia. **Results:** 10 (32%) were diagnosed with EoE. Male gender was more common in the EoE group: 8 vs 2 (80% vs 20%). There was no difference in family history of atopy, EGA, mode of delivery, use of breast milk or presence of gastrostomy. Tracheomalacia affected 9 of 21 (48%) non-EoE vs 7 of 10 (70%) EoE; aspiration pneumonia 67% non-EoE vs 30% EoE. Age of initial dilation was later in EoE and 25% were >2y at first dilation. Inhaled steroid exposure under 2y of age was higher in the non-EoE group 10 of 19 (52%) vs 1/10 (10%). Steroid use at time of endoscopy was low in both groups (24% vs 22%) so unlikely to have a treatment effect for EoE.

Conclusion: 32% of our EA subjects had a diagnosis of EoE. Male sex was a risk factor for EoE but there was no difference in family history of atopy. Early exposure to topical steroid was more common in the non-EoE group. Whether early topical steroid is somehow protective from EoE is not clear, as our numbers are small, but this observation needs further follow up.

P-39

ENDOSCOPIC MULTIDISCIPLINARY TEAM FOR EA-TEF PATIENTS : A PILOT STUDY

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Multidisciplinary speciality clinics has been shown to improve clinical outcomes in a variety of conditions such as esophageal atresia- tracheo-esophageal fistula(EA-TEF)patients.

Goal : To review the outcomes of 1 year of combined multidisciplinary endoscopic procedures (gastroscopy,rigid bronchoscopy, flexible bronchoscopy and/or methylene blue test) on patients with EA-TEF to demonstrate that we can extend the benefits multidisciplinary teams to the endoscopic procedures.

Methods : All multidisciplinary endoscopic procedures on patients with EA-TEF between March 2013 and June 2014 were reviewed.

Results: 25 procedures were performed at Montreal Children's Hospital on 21 patients with EA-TEF aged 7 months to 17 years.Two patients had 2 procedures and one had 3 procedures. 11 procedures consisted of : gastroscopy(EGD), flexible and rigid bronchoscopies and a methylene blue test. 7 procedures consisted of an EGD with rigid bronchoscopy. Indications for various procedures were tailored to each child.Airway evaluation as well as excluding a laryngeal cleft or recurrence of TEF accounted for the majority of Respirology and Otolaryngology procedures.6 procedures were performed to cure a recurrent TEF. Gastroenterology procedures included screening esophagitis as well as endoscopic treatment of esophageal stricture. The common approach allowed the diagnosis of 5 new esophagitis,1 esophageal stricture,15 tracheomalacia and 8 laryngeal clefts.We also confirmed the existence of 2 laryngeal clefts, 4 recurrent TEF and 2 esophageal strictures.Six procedures led directly to further investigations (CT-scan or thoracic MRA) before a treatment could be done.

Conclusion : A multidisciplinary approach is clearly advantageous to address the medical and surgical needs of the EA-TEF population.

P-40

RADIATION EXPOSURE AND ATTRIBUTABLE LIFETIME CANCER MORTALITY RISK FOR PATIENTS WITH ESOPHAGEAL ATRESIA

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Children with esophageal atresia (EA) undergo considerable amounts of diagnostic imaging and consequent radiation exposure throughout their course. We evaluated the radiological procedures performed on EA patients and the cumulative radiation exposure and attributable cancer risk.

Methods

With IRB approval, patients with EA managed from 2001-2013 were investigated. Demographics, EA subtype and number and type of radiological investigations were gathered. Existing normative data was used to estimate the cumulative radiation exposure and lifetime cancer risk per patient.

Results

There were 72 children, 57 with type C atresia. Median follow-up was 5.7 years (mean=6.33+/-3.9). Table 1 demonstrates the amount of imaging and radiation dose during admission and through follow-up, with an overall median 6.47 mSv/patient. This represents 3 times the annual total normative radiation dose. Radiation exposure in the neonatal period was 5.7 mSV, correlating with an estimated cumulative lifetime mortality risk of cancer of 1:200 per infant.

Conclusion

Children with EA are exposed to significant amounts of radiation during hospitalization and throughout follow-up, which may contribute to the documented increase in esophageal cancer rates in these patients. Elimination of superfluous imaging appears warranted, as does direct dosimetry measurements for at-risk patients.

Imaging	During Admission Median (mean +/- SD)	During Follow up Median (mean +/- SD)	Total Median (mean +/- SD)	Estimated effective dose/study (mSv)	Total Median effective dose combined (mSv)
Plain CXR	7 (13.2+/- 19.4)	4 (9.13 +/- 12.2)	17 (22.3 +/- 24.4)	0.02*	0.34
Plain AXR	2 (3.6 +/- 5.3)	0 (1.49 +/-3.5)	2 (5.1 +/- 7.2)	0.02*	0.04
UGI	1 (0.88 +/- 1)	0 (0.68 +/- 0.9)	1 (1.6 +/- 1.4)	5.44**	5.44
Other Fluoro	0 (0.9 +/-1.2)	1 (1.37+/-1.6)	1 (2.3+/- 2.6)	0.65**	0.65

*From Puch-Kapst et al. 2009 **From Dimitradis et al. 2011

Summary of radiology and effective dose administration for cohort of patients with esophageal atresia

P-41

WHAT MOTHERS THINK OF COMMUNICATION ABOUT THEIR CHILD WITH TRACHEOESOPHAGEAL FISTULA (TEF)

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Introduction: Tracheoesophageal Fistula (TEF) is a congenital anomaly that requires complex initial treatment. Children with TEF face multiple challenges including: surgeries, hospital stays, and complications. These children require numerous health care services to manage their complicated condition. This is a challenging time for the parents of a child with TEF. A multidisciplinary clinic was started in 2006 to coordinate the complex care of these children and their families.

Aim: To gain an understanding of parental perceptions of the communication provided through this multidisciplinary clinic.

Methods: Interviews were conducted with three primary caregivers of children with TEF born between 2010 and 2011. Qualitative data obtained from these interviews was analyzed to identify whether the TEF clinic has contributed to improvement of care.

Results: Parent interviews include positive feedback for these themes: appropriate information, coordination of care, positive relationships, and anticipatory guidance. Improvements could be made to coordination of care between disciplines especially during pre-surgical communications.

Conclusions: A multidisciplinary approach to complex care of children with TEF is beneficial. There was positive feedback from parents with children enrolled in this clinic, indicating successful aspects of the clinic care and communication. Some aspects of care remains fragmented, and further aims to improve the TEF Clinic are required.

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THE INFLUENCE OF CONGENITAL HEART DEFECTS ON MID TERM OUTCOMES OF ESOPHAGEAL ATRESIA PATIENTS

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Introduction and aim

Major congenital heart defects (CHD) represent a significant risk factor predicting mortality in esophageal atresia (EA) infants. However, the influence of CHD in post-postoperative outcomes of patients with EA is poorly explored. **Aim** of the present study is to evaluate outcomes at mid term follow-up in AE patients in respect of CHD.

Methods

Retrospective analysis of all EA infants treated between January 2008 and December 2012 was performed. Patients were divided into two groups based on the presence of major CHD (requiring cardiac surgery). Mid term outcomes were compared between the two groups. Statistical analysis was performed with Fisher exact test and Mann-Whitney test, as appropriate ($p < 0.05$ was considered significant).

Main Results

During the study period 76 patients were prospectively treated and enrolled into our follow-up program. Mean follow-up period was 3,2 y (1-6,1y). Table 1 summarized main results.

Conclusion

Patients affected by EA and major CHD, even if experienced a more severe perinatal period, presented less esophageal stenosis requiring dilatations and re-do surgery in comparison with non-CHD EA infants. Since it is demonstrated that major CHD depends on collagen defects, this could explain the major "plasticity" of the esophagus in EA-CHD infants.

	Non-CHD 55	CHD 21	p
Gestational age, weeks (IQR)	38 (36-39)	37 (34.25-38)	0.05
Birth weight, gr (IQR)	2725 (2213-3045)	2500 (2250-2900)	0.4
Type A/B vs C/D	5/2 vs 48/0	2/0 vs 18/1	1
Long gap EA (%)	11/55 (20)	6/21 (28)	0.55
Cervical esophagostomy (%)	7 (13)	2 (10)	1
Referral	8 (15)	3 (14)	1
Primary anastomosis	43 (78)	17 (81)	1
Major Leak	6 (11)	0 (0)	0.12
Dilatations, median (IQR)	2 (0-3)	1 (0-1)	0.01
Need for esophageal stent (%)	3 (6)	1 (5)	1
Antireflux surgery GERD (%)	8 (15)	3 (14)	1
Late deaths (%)	2 (4)	1 (5)	1
Surgery related deaths (%)	0	0	1

P-43

LONG-TERM HEALTH-RELATED QUALITY OF LIFE AFTER COMPLEX AND/OR COMPLICATED ESOPHAGEAL ATRESIA IN ADULTS AND CHILDREN REGISTERED IN A GERMAN PATIENT SUPPORT GROUP

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Introduction

Health-related quality of life (HRQoL) after esophageal atresia (EA) repair is postulated to be good. However, little is known about the long-term results after repair of complex and/or complicated EA regarding HRQoL.

Aim

Aim of this study was to investigate long-term HRQoL after delayed anastomosis, esophageal replacement, major revisions, or multiple dilatations in patients registered in a support group.

Methods

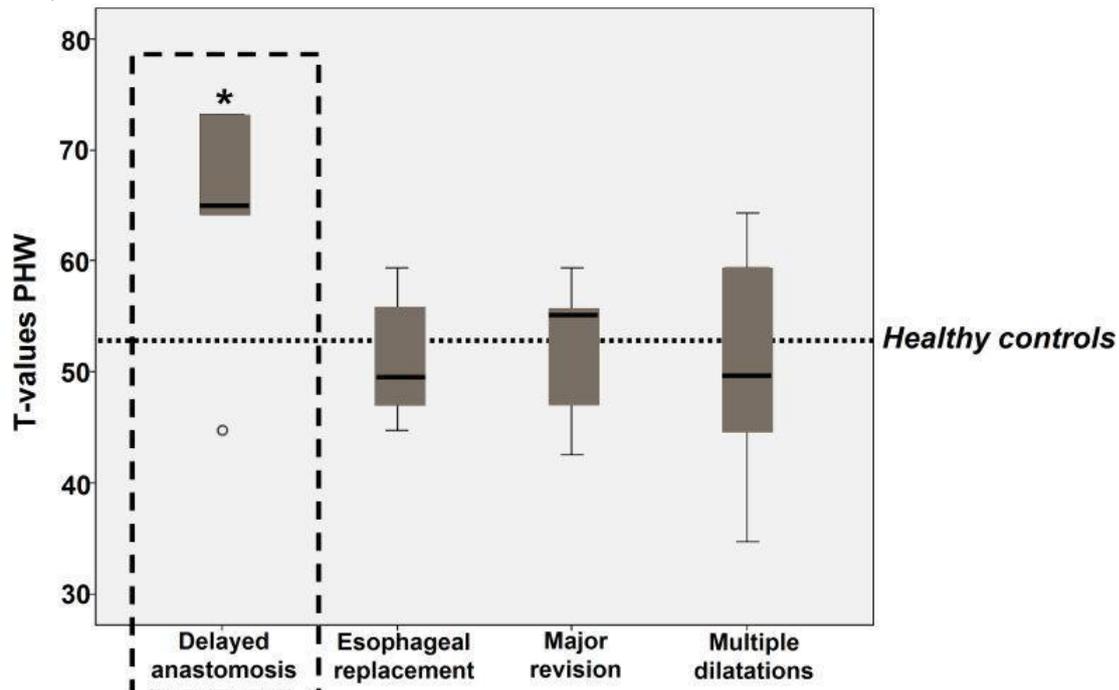
Patients registered in the German patient support group database (KEKS) were enrolled and allocated to subgroups according to surgical treatment and age. HRQoL was evaluated using validated questionnaires (GIQLI, WHO-5, KIDSCREEN27).

Results

Complete follow-up (mean 14.5 ± 9.8 years) was available for 90/92 patients. Patients were allocated to subgroups *delayed anastomosis* (n=28), *esophageal replacement* (n=27), *major revisions* (n=15), and *multiple dilatations* (n=20). Adult patients presented with impaired well-being according to WHO-score and gastrointestinal function (GIQLI). In contrast, HRQoL of children was comparable to controls in most KIDSCREEN27-dimensions. Delayed anastomosis was associated with most-favourable HRQoL [Figure 1]. Regarding physical well-being (PHW), these children scored significantly better than controls [64.01 ± 10.40 vs. 52.36 ± 8.73 ; $p=0.0011$], children after replacement [51.40 ± 5.70 ; $p=0.008$], revisions [52.04 ± 6.97 ; $p=0.026$], and multiple dilatations [50.22 ± 9.67 ; $p=0.04$].

Conclusion

HRQoL after complex and/or complicated EA is excellent in children registered in a patient support group. In adults, disease-specific symptoms negatively affect HRQoL. Our data indicate that saving the esophagus may achieve the best HRQoL.



KIDSCREEN27 score given for the dimension Physical Well-Being (PHW).

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RADIOFREQUENCY ABLATION IN TREATMENT OF BARRETT'S ESOPHAGUS ALSO IN ESOPHAGEAL ATRESIA

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Barrett's esophagus (BE) is rare in children but associated to long-standing GERD caused by esophageal atresia, peptic strictures, caustic esophagitis. Radiofrequency ablation (RFA) with proton pump inhibitor (PPI) is the new treatment proposed for adult BE, with known safety and effectiveness. No series are published until now in BE with pediatric onset.

Methods

After symptoms evaluation, endoscopy-histology assessment of BE, pH monitoring, we performed HALO(360°) RFA followed by twice daily PPI and 3-monthly surveillance for up to 12 months. If metaplasia or dysplasia were present at follow-up, the patients received other ablations.

Results

Three patients (males; BE at 12, 13 y, 14 y) were treated; one with low-grade dysplasia (27 y) C4 M8, esophageal atresia repaired during the first year of life, and two with non-dysplastic intestinal metaplasia (20 and 30 y), one C4 M8 and one C8 M10. The mean baseline BE length was 9 cm; all patients had a hiatal hernia undergone a previous fundoplication, two of which after onset of peptic stricture treated with semirigid dilations and PPI; pH-monitoring was normal. Two, three and four sessions were performed, with regression of metaplastic-dysplastic tissue up to 90% of the treated surface, at a mean follow up of 6.3 months (range 2-10 mths). Quality of life of all patients already improved, after the first ablation.

Conclusion

RF ablation could represent a new promising opportunity of managing BE in young patients with precocious onset of metaplasia/dysplasia, to be confirmed in long term perspective studies.